The Journal of
Bone and Joint Surgery

EDITORIALS AND ANNOTATIONS

GENETICS IN ORTHOPAEDICS

It is a truism today that direct environmental causes of disease and malformation, such as infection and malnutrition, though not trauma, are being overcome. In consequence, an increasing proportion of disease and malformation is genetically or in part genetically determined. Thus rickets in this country is now seldom due to simple nutritional vitamin D deficiency, but usually a manifestation of one of the genetically determined abnormalities affecting calcium and phosphate metabolism. Similarly, the problems of muscle weakness and imbalance from poliomyelitis will disappear, but those associated with the muscle dystrophies or with spina bifida cystica remain and, with a greater salvage of malformed children, may even increase.

In the past there has been an unfortunate tendency to regard genetically determined disorders as untreatable. This is by no means always so. Fully effective treatment, involving perhaps the recognition and then the replacement of a missing enzyme, may be many years ahead. Remedial measures, however, will often be possible, though requiring skill and determination in their application.

A study of the genetics of a condition can contribute towards the handling of genetically determined disorders in various ways. When the genetics of the condition are known it is possible to advise an affected individual accurately on the risk of transmitting the condition to his child, or to advise normal parents who have had an affected child on the risk of having a second affected child. In addition, knowledge that a child is at risk may lead to early diagnosis and treatment—as for example the diagnosis and then maintenance therapy with large doses of vitamin D before the onset of skeletal deformity in affected children of men and women with dominant primary vitamin D resistant rickets (Dent and Harris 1956; Linneroth, Hallgren, Palmen and Zetterström 1958).

Genetic knowledge, too, may give increased precision in diagnosis. Studies of several affected members of a family may show that a mild and severe form of a disorder have essentially the same etiology. In this issue Lennon, Schechter and Hornabrook show how family studies have led to the recognition of varied manifestations of Engelmann's disease. Conversely, genetic analysis may show that there are two distinct forms of a disease which had been regarded as one. The autosomal and the sex-linked recessive forms of gargoylism were separated genetically before clinical distinction was made between them. In the same way, a form of osteochondrodystrophy with some similarities to, but less severe than, the autosomal recessive form of Morquio's disease has been identified by its sex-linked recessive pattern of inheritance (Maroteaux, Lamy and Bernard 1957).

It must be recognised, however, that the genetics of many orthopaedic conditions are still unexplored. The methods of genetic investigation include twin studies, family studies, cytological studies and studies of similar conditions in animals. Reports of single twin pairs, such as that of Hull in this issue, are not without value; but for an estimate of the general importance of genetic factors for a particular condition a large unselected series of co-twins of index patients is needed. It is the percentage of identical co-twins similarly affected, in

comparison with the percentage of like-sex fraternal twins similarly affected, that is informative. This cannot be established satisfactorily by summarising the twin pairs reported in the literature, since various forms of bias influence an author's decision whether or not to report a particular pair. Idelberger's (1939, 1951) twin studies of talipes equinovarus and congenital dislocation of the hip are a model of what can be done in assembling and investigating a unselected series. Twin studies, however, give little information about the particular type of genetic factors involved. For this, family studies are needed.

Individual family studies, such as those of Engelmann's disease and osteochondritis dissecans reported in this issue by Lennon and his co-authors and by Stougaard, are valuable provided that, as in these studies, the relatives are carefully examined. A report of a family, with for example two brothers and a sister affected, is of little help in elucidating genetic mechanisms unless one knows for certain whether or not the parents are affected. As with twin studies the most informative family studies are those based on a large consecutive series of index patients. Good examples are the surveys of achondroplasia in Denmark by Mørch (1941), of childhood muscular dystrophy in Yorkshire by Blyth and Pugh (1959), and of both these conditions in Ulster by Stevenson (1957). From such studies it is possible to get an idea of the types of genetic factors involved, the relative frequency of each type, the range of clinical effects they may produce, and whether there is a residue of patients for which one cannot account by small family size or fresh mutation, and for which some non-genetic explanation must be sought.

Cytological studies with direct microscopic examination of human chromosomes in tissue culture of skin, marrow or blood, are a new development. Experience hitherto suggests that major chromosome anomalies produce skeletal abnormalities, but as part only of a complex syndrome. Examples are the brachycephaly, hypotonia, joint laxity and altered acetabular slope in mongolism (due to an extra chromosome 21), and the dwarfism, webbed neck and increased carrying angle of the arm in children with Turner's syndrome (from absence of a second sex chromosome). It is to be expected that minor chromosome abnormalities will also become detectable and that the effect of some of these will be primarily on the skeleton.

The search for, and the analysis of, disorders in animals apparently similar to human conditions has its advantages, and also its dangers. The advantages are that by controlled breeding it is possible to study the genetic mechanisms in detail. For example two animals, each heterozygous for the same dominant gene, may be mated to see the effect of the gene in the animals homozygous for it. This will often produce an extreme manifestation of the disorder. It is also possible to kill the pregnant animal and study the development of the disorder in the foetal animal. The study by Berry, reported in this number, of a genetically determined maldevelopment of the nucleus pulposus, and its relation to prolapse of the disc in the mouse is a good example of what can be done. But there must always be care in applying conclusions drawn from disease in animals to man. Only perhaps when the biochemical deficiency is shown to be identical in the two species can one be satisfied that the two conditions are essentially the same.

The two branches of medicine in which genetic studies have been most extensive are dermatology and ophthalmology, for the good reason that the bodily systems involved may be examined directly. Radiography and biopsy offer much the same facilities for the study of orthopaedic conditions, and we may hope for considerable advances in the knowledge of genetically and part-genetically determined orthopaedic disorders. The most rewarding developments will be, perhaps, in the disorders with mixed genetic and environmental causation, such as spina bifida cystica, congenital dislocation of the hip, club-foot, scoliosis and osteoarthritis. Genetic analysis for these conditions is more difficult than for those essentially due to single gene abnormalities; but they are relatively common, and their mixed etiology offers the prospect of a reduction in their incidence by special precautions applied to those who are genetically predisposed to develop them.

C. O. CARTER.

REFERENCES

BLYTH, H., and PUGH, R. (1959): Muscular Dystrophy in Childhood. The Genetic Aspect. *Annals of Human Genetics*, **23,** 127.

DENT, C. E., and HARRIS, H. (1956): Hereditary Forms of Rickets and Osteomalacia. *Journal of Bone and Joint Surgery*, **38–B,** 204.

DELBERGER, K. (1939): Die Ergebnisse der Zwillingsforschung beim angeborenen Klumpfuss. *Verhandlungen der Deutschen Orthopädischen Gesellschaft*, **33,** 272.

DELBERGER, K. (1951): *Die Erbpathologie der sogenannten angeborenen Hüftverrenkung.* Munich: Urban und Schwarzenberg.

INNEROTH, K., HALLGREN, B., PALMEN, J., and ZETTERSTRÖM, R. (1958): Primary (Genuine) Vitamin D Resistant Rickets. *Acta Paediatrica*, **47,** 568.

MAROTEAUX, P., LAMY, M., and BERNARD, J. (1957): La dysplasie spondylo-epiphysaire tardive. *Presse Médicale*, **65,** 1,205.

MØRCH, E. T. (1941): *Chondrodystrophic Dwarfs in Denmark.* Copenhagen: Ejnar Munksgaard.

STEVENSON, A. C. (1953): Muscular Dystrophy in Northern Ireland. *Annals of Eugenics*, **18,** 50.

STEVENSON, A. C. (1957): Achondroplàsia: an Account of the Condition in Northern Ireland. *American Journal of Human Genetics*, **9,** 81.

NEUROPATHIC JOINTS

Charcot's original paper of 1868, " On some arthropathies which appear to depend on a lesion of the brain or spinal cord," described two forms of joint lesions associated with disease of the nervous system.

The first was that occurring in the course of locomotor ataxia giving rise to swelling, usually of the knee joint, of sudden onset, in the absence of fever, redness and pain, and accompanying excessive mobility of the joint. The second affected the joints of paralysed limbs in hemiplegia of cerebral origin, and was accompanied by severe pain on movement with redness, swelling and warmth of the affected joints. Only the former retains the eponym Charcot's joint. As the years passed, similar joint changes were found in other diseases; for example, syringomyelia, affecting especially the elbow and shoulder; diabetes mellitus, especially in the ankle and foot, though here both ischaemia and infection played a part; and many other spinal cord, posterior root and peripheral nerve lesions which resulted in analgesia, and often in loss of proprioception. For all of these many modern authors use the generic term *neuropathic joint.*

Charcot held the view that the joint changes resulted from a removal of the " trophic " influence of the nervous system over bones and joints, but this was challenged by Westphal (1881) with the strong support of Volkmann (1882), and later of Rotter (1887). Volkmann related the destruction of the joint to repeated trauma which, owing to the analgesia, was unperceived, and owing to the loss of proprioception was unmitigated when the joint was exposed to extreme movements; hence repeated damage to the joint resulted in the classical clinical and radiological picture (Cohen 1955).

Volkmann's view of the pathogenesis of a Charcot's joint received strong support from the experimental work of Eloesser (1917) confirmed by Delano (1946). Eloesser showed that a Charcot joint in cats, although it may rarely follow simple section of the posterior nerve roots, consistently follows that operation if concomitant joint injury is inflicted. Subsequent observations in man (Casagrande, Austin and Indeck 1951) have tended to confirm that not only must there be loss of pain, but also proprioceptive loss and continued use of the joint.

In recent years two conditions have been added to the list of causes of Charcot's joint. First is the fascinating condition of congenital universal indifference to pain, of which van der Houwen describes an example in this issue of the Journal. Secondly, there is the arthropathy following intra-articular corticosteroid therapy.

CONGENITAL UNIVERSAL INDIFFERENCE TO PAIN

In 1938 Ford and Wilkins described three children between the ages of seven and nine who could not recognise any painful stimulus. This was a condition that had to be distinguished from the absence of reaction to pain which is occasionally seen, for example, in hysteria, hypnosis and those depressive psychoses in which self mutilation is not uncommon. It is, of course, within most people's experience that in extreme excitement, anger or fear, as in combat or sport, they may not feel pain or react to it.

In true congenital indifference to pain there are several criteria. First, the condition dates from birth or early life and is permanent. Jewesbury's report (1951) of a boy who at sixteen months appeared grossly indifferent to pain but was normal at two and a half years should be excluded from this group. Secondly, analgesia is found throughout the whole body, and to all forms of painful stimuli—prick, pinch, burn, intense cold, faradic shock, etc.; and the normal physiological phenomena that accompany pain, such as changes in pulse rate and blood pressure, are absent. Accompanying slight defects for other forms of sensation have occasionally been described, but there is never any gross sensory loss except for pain.

In a few patients evidence of mental subnormality and other neurological defects have been noted, but in most there is normal or accelerated intellectual development. This adds weight to the view that we are dealing with " insensitivity " and not " indifference " to pain.

The condition is brought to light by incidents which unmask this insensitivity to pain, such as bruises, cuts, burns or dental extractions; so it is not surprising that many patients with congenital indifference to pain have presented with bone or joint lesions. These include fractures, bone atrophy, osteomyelitis, joint luxation and arthropathies of the Charcot type, such as that which van der Houwen describes.

One of the most striking of these patients has been the subject of four reports in the literature, including that by Petrie (1953), who described the orthopaedic complications of this patient, and Feindel (1953), who examined specimens of skin, fascia, periosteum, bone and cartilage, and found nerve fibres in all. He concluded from his examination that " from the histological evidence available, it seems probable that the defect in perception of pain, and in the reflex productive effects of pain in this subject lies in central rather than in peripheral nerve structures." Since van der Houwen wrote his paper this patient has died, and a full account of her history and necropsy has been published by Baxter and Olszewski (1960). They carried out a complete examination of the nervous system but " failed to demonstrate any anatomical abnormality of those nervous system structures thought to be concerned with the transmission, elaboration and perception of pain impulses," though they could not " exclude the possibility that the defect is actually an anatomical one, either submicroscopic or in terms of organisation rather than structure."

INTRA-ARTICULAR CORTISONE

The method of treating an arthritic joint by intra-articular injections of cortisone was introduced by Hollander in 1951. Ten years later he was able to report his experience and that of his colleagues of 100,000 injections in 4,000 patients, and claimed that instability of a weight-bearing joint occurred in only thirty-seven joints in twenty-six patients (0·7 per cent). In four joints, however, there was an extensive absorption of bone associated with aseptic necrosis, and though they claimed that there was no loss of sensation or proprioception in these unstable joints, they did speak of them as " relatively painless."

Other authors have reported rapid deterioration of joints, especially of the osteoarthritic hip, after repeated hydrocortisone injections, leading to a picture which is clinically and radiologically indistinguishable from a Charcot joint (Chandler, Jones, Wright and Hartfall 1959).

Two factors in pathogenesis are clearly: 1) the relief of pain which corticosteroids give and 2) the consequent greater mobility and use of the joint with increasing damage. A third

factor is almost certainly the softening of bone which high concentrations of corticosteroids produce in their neighbourhood, greater than, but akin to, the osteoporosis which often follows corticosteroids by mouth.

It is clearly imperative that careful radiological supervision should control intra-articular therapy with corticosteroids.

The story of Charcot's joints illustrates three stages in the evolution of our knowledge of any clinical phenomenon. In the first it is regarded as a pathognomic sign of the disease in which it was described. Later it is found to occur in other diseases, sometimes of known cause. Finally its pathogenesis is understood in terms of dysfunction for which an anatomical substratum might be revealed. It is only at this final stage that we can claim to understand why in newly described diseases old signs reappear. COHEN OF BIRKENHEAD.

REFERENCES

BAXTER, D. W., and OLSZEWSKI, J. (1960): Congenital Universal Insensitivity to Pain. *Brain*, **83,** 381.

CASAGRANDE, P. A., AUSTIN, B. P., and INDECK, W. (1951): Denervation of the Ankle Joint. *Journal of Bone and Joint Surgery*, **33–A,** 723.

CHANDLER, G. N., JONES, D. T., WRIGHT, V., and HARTFALL, S. J. (1959): Charcot's Arthropathy Following Intra-articular Hydrocortisone. *British Medical Journal*, **i,** 952.

CHARCOT, J. M. (1868): Sur quelques arthropathies qui paraissent dépendre d'une lésion du cerveau ou de la moëlle épinière. *Archives de Physiologie Normale et Pathologique*, **I,** 161, 379.

COHEN, Sir Henry (1955): In *Textbook of the Rheumatic Diseases*. Edited by W. S. C. Copeman. Second edition, p. 282. Edinburgh and London: E. & S. Livingstone Ltd.

DELANO, P. J. (1946): The Pathogenesis of Charcot's Joints. *American Journal of Roentgenology*, **56,** 189.

ELOESSER, L. (1917): On the Nature of Neuropathic Affections of the Joints. *Annals of Surgery*, **66,** 201.

FEINDEL, W. (1953): Note on the Nerve Endings in a Subject with Arthropathy and Congenital Absence of Pain. *Journal of Bone and Joint Surgery*, **35–B,** 402.

FORD, F. R., and WILKINS, L. (1938): Congenital Universal Insensitiveness to Pain. *Johns Hopkins Hospital Bulletin*, **62,** 448.

HOLLANDER, J. L., BROWN, E. M., Jun., JESSAR, R. A., and BROWN, C. Y. (1951): Hydrocortisone and Cortisone Injected into Arthritic Joints. *Journal of the American Medical Association*, **147,** 1629.

HOLLANDER, J. L., JESSAR, R. A., and BROWN, E. M., Jun. (1961): Intra-synovial Corticosteroid Therapy: a Decade of Use. *Bulletin on Rheumatic Diseases*, **11,** No. 5.

JEWESBURY, E. C. O. (1951): Insensitivity to Pain. *Brain*, **74,** 336.

PETRIE, J. G. (1953): A Case of Progressive Joint Disorders Caused by Insensitivity to Pain. *Journal of Bone and Joint Surgery*, **35–B,** 399.

ROTTER, J. (1887): Die Arthropathien bei Tabiden. *Archiv für Klinische Chirurgie*, **36,** 1.

VOLKMANN, R. von (1882): Comment on Westphal's paper. *Zentralblatt für Chirurgie*, **9,** 174.

WESTPHAL, C. (1881): Gelenkerkrankungen bei Tabes. *Berliner Klinische Wochenschrift*, **18,** 413.

ELECTRODIAGNOSIS*

C. B. WYNN PARRY, ROYAL AIR FORCE

Nearly eighty years ago Erb (1883) described the faradic-galvanic test and thus laid the foundation of electrodiagnosis in lower motor neurone lesions. Notable advances made in electrodiagnosis in the last fifteen years are reviewed in this paper, which will also discuss the indications, limitations and usefulness to the orthopaedic surgeon of the various techniques.

Electrodiagnosis has five main uses: to detect denervation; to detect reinnervation before the clinical signs are apparent; to assess the amount of denervation in a muscle; to assess the progress of a lesion—whether degenerating or recovering; and to find out if the lesion is in the anterior horn cell, the peripheral axon, the neuromuscular junction or the muscle itself.

Up to a few years ago the universal method of electrodiagnosis was the faradic-galvanic test. As this test is still used quite widely and as it is now realised that it can often be fallacious the test will be described and its shortcomings analysed in detail.

The response of a normally innervated muscle to a short duration low voltage (or faradic) current is a brisk response. The response to a long duration (galvanic) current is also brisk. When a muscle is denervated the faradic response is said to disappear and the response to the galvanic current is said to be sluggish.

Although Erb specifically stated in his book that a denervated muscle can be made to contract with a faradic current, it is widely believed that if a muscle responds to faradism it shows that some degree of innervation is present. It has been shown (Wynn Parry 1954) that over 75 per cent of muscles known to be totally denervated respond to a short duration current using an electronic stimulator.

Ritchie (1954) analysed the results of over 10,000 faradic-galvanic tests done during the last war on injured peripheral nerves, and related these to the operative findings and the clinical progress. He found that the test had only a 50 per cent chance of being right. Paradoxically, he showed that the better the physiotherapist the more likely was the result to be misleading. The reason for this was that the good physiotherapist would encourage the patient to take a strong current and thus to make more efficient use of the apparatus which was, in those days, a faradic coil. This coil was unreliable and the current produced varied with the mains voltage, which might fluctuate, and with the state of the contacts; moveover the current produced varied considerably between one instrument and another. Clearly the higher the output of a particular machine and the better the condition in which it was kept, the more likely it was to produce a current strong enough to give a positive faradic response in a denervated muscle which will often respond to a short duration current with a high voltage. This in itself was a perfectly correct observation, but the interpretation was wrong, for a response to faradism does not always mean the presence of recovery.

The modern electronic stimulators used in treatment give a high output and are much more comfortable for the patient as they provide a square wave stimulus instead of a condenser discharge. Therefore faradic-galvanic tests done with these machines and the results interpreted in the conventional manner will be even more misleading.

It will be shown later that the faradic test is really only detecting two points on a full strength duration curve and, apart from the drawbacks already described, it is only qualitative; comparisons from week to week to assess progress are, therefore, impossible.

On the other hand the plotting of a strength duration (SD) curve is simple, extremely reliable, gives an accurate quantitative assessment of the state of innervation and takes less time than the faradic-galvanic test. Because of this the latter is obsolete and should never be used.

* This paper was read at the British Orthopaedic Association meeting in Oxford, October 2–4, 1958.

STRENGTH DURATION CURVES

Nerve and muscle are both excitable tissues because they have polarised membranes, and both have a characteristic response to electric currents. This response can be measured by strength duration curves which also allow the proportion of normally innervated to denervated fibres to be assessed.

The muscle is stimulated with an electronic stimulator. This gives a series of square waves, the longest duration being 300 milliseconds and the shortest 0·01 milliseconds.

The amount of current needed to cause just perceptible contraction to the naked eye is measured at the longest pulse duration, which is 300 milliseconds. This is the rheobase which, by definition, is the amount of current flowing for infinite time required to cause a just perceptible contraction. Experience has shown that a time longer than 300 milliseconds will never lower the current needed to produce the smallest contraction. The rheobase is thus a measure of the electrical threshold of the tissue. Below this threshold, however long the current lasts, it is not possible to cause the muscle to contract.

In the same way, the amount of current needed to produce a just perceptible contraction at each of a series of progressively shorter pulse durations—100, 30, 10, 3, 1, 0·3, 0·1, 0·03 and 0·01 milliseconds—is measured, and the curve relating strength of current with duration of current is drawn and is called the strength duration (or intensity duration) curve. The actual test takes no more than two to three minutes.

Technique of making a strength duration curve—The detailed technique of plotting a strength duration curve is as follows: the part should be in a good light and reasonably warm. In cold weather ten minutes' exposure to a radiant heat lamp is advisable. For a longitudinal technique, two one-centimetre brass electrodes covered in lint well soaked in weak warm saline solution are laid gently on the muscle in its longitudinal plane on either side and equidistant from the point across which the best contraction is produced. The exact placing of electrodes varies according to the size of the muscle. For the small muscles of the hand the electrodes are applied at either end of the muscle, for larger muscles they will be some inches apart; with the quadriceps, for example, the electrodes are four inches apart. Care is taken that there is very little pressure from the electrodes on the muscle, for even slight pressure can conceal from sight a very slight contraction.

The longitudinal reaction is preferable to an indifferent electrode on the spine and an active electrode over the motor point, because, with the latter method, spread of the current to adjoining muscles can be very confusing, particularly when they are normal and the muscle being tested is denervated or partially innervated. With the longitudinal reaction, the current can be concentrated between the electrodes in the plane of the muscle and spread of the current is much less troublesome.

The pulse duration selector switch is set at 300 milliseconds, the pulse repetition frequency to 0·5 seconds (one impulse every two seconds) and the apparatus is checked to ensure accurate calibration. The voltage is then gradually increased until a good contraction of the muscle is observed and the quality of the contraction—whether brisk or sluggish—is noted. Six to eight strong contractions are produced, as this is a useful final precaution in lowering skin resistance, and the rheobase will then be the same at the end of the test as at the beginning. The voltage is then gradually lowered until the contraction disappears. The voltage is increased again a fraction at a time until the contraction just appears. This voltage is read off the dial and recorded, and is, by definition, the rheobase. With electronic stimulators, the end point for the rheobase is highly critical; lowering the intensity by 1 volt causes the contraction to disappear. The pulse duration switch is then moved to the 100 millisecond position, without altering the voltage or the position of the electrodes, and the muscle again observed. If the contraction has disappeared the voltage is increased until it reappears and is then slowly reduced until it again disappears. Once more the voltage is increased until the minimal contraction is seen and the reading on the voltage dial recorded. The voltage required to produce minimal contraction at the remaining eight pulse durations is determined in the same manner. A curve is now drawn relating pulse duration with voltage—the so-called strength duration curve.

In the majority of normal muscles a response is found with the shortest pulse durations. In denervated muscles it is found that at the shorter pulse durations no contraction is seen with the maximum output of 200 volts. It is clear that when there is no response with 200 volts at, say, 0·3 milliseconds, there is no need to test the muscle at shorter pulse durations and there will be only six points for the curve.

It is more accurate for the examiner who is testing the muscle both to manipulate the switches and voltage dial and to observe the contractions. It is of great convenience to have an assistant to record the figures as they are obtained.

Where muscles are inserted by easily palpable tendons, it has been found more accurate to feel for tendon movement as well as to watch for the contraction between the electrodes. Particularly with the dorsiflexors of the ankle, no contraction may be visible yet a movement of the tendon is palpable.

On completion of the test the voltage dial is returned to zero before returning the pulse duration switch back to 300 milliseconds. Omission of this precaution has, on occasions, led to a current of 200 volts of 300 milliseconds duration passing through the examiner if he is holding the active end of the electrodes, which, if not dangerous, is extremely unpleasant.

Very few patients have found the test uncomfortable although some patients find the 300 millisecond pulse, particularly with a high rheobase, rather a sharp and unpleasant sensation; for

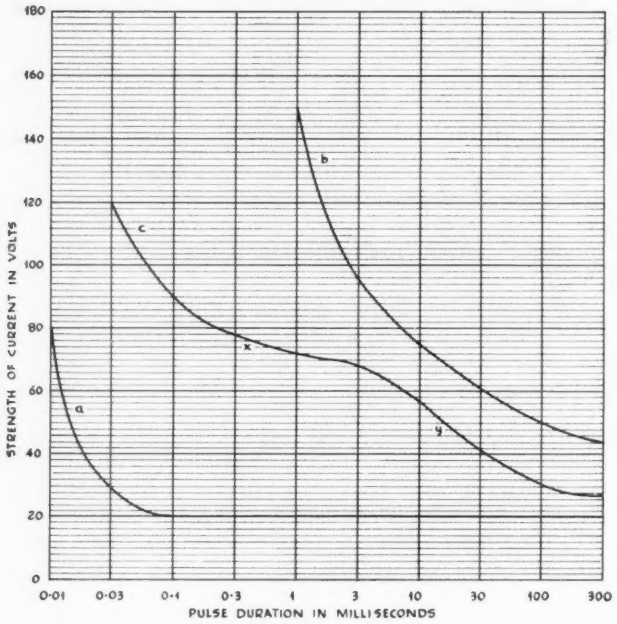


FIG. 1

Strength duration curves on (a) normal, (b) denervated, and (c) partially innervated muscle. That part of curve (c) marked x represents the innervated component (response of nerve) and that part marked y represents the denervated component (response of muscle).

these the rheobase is taken at 100 milliseconds instead. At the shortest pulse durations, even 200 volts is barely perceptible. It is very rare that a test has to be stopped because of discomfort to the patient.

On an abnormal muscle the average time for performing a strength duration curve is four minutes. A curve on a normal muscle may take as little as fifty seconds to perform.

The characteristic curves of normally innervated, denervated and partially denervated muscle are shown in Figure 1. " a " is the curve of a normally innervated muscle. It is the excitability response of nerve, for when a normal muscle is electrically stimulated the current affects the intramuscular nerve fibres as these are much more excitable than muscle fibres. " b " is the curve of a completely denervated muscle. Here the current can only affect the muscle fibres, and so this gives the response of muscle. The curve rises steeply with shorter duration impulses, and there is no response at the short durations. If, however, this curve were prolonged to the right it would be horizontal, for the value of current to produce a just

perceptible contraction in 300 milliseconds is, by definition, the rheobase. Comparison, therefore, of these two curves shows that they are of the same shape but that of muscle is shifted to the right indicating a lower excitability than nerve. "c" is the curve of a partially denervated muscle. The portion marked x represents the innervated component or the response of nerve, for it corresponds to curve "a." The portion marked y represents the denervated component—or the response of muscle—for it corresponds to curve "b." Thus the SD curve allows one to assess the ratio of muscle to nerve components—that is the ratio of denervated to innervated fibres, and to some extent this can be a quantitative assessment. In Figure 1 curve "c" is S-shaped and suggests that the ratio is 1 : 1, that is, that there is 50 per cent of denervation.

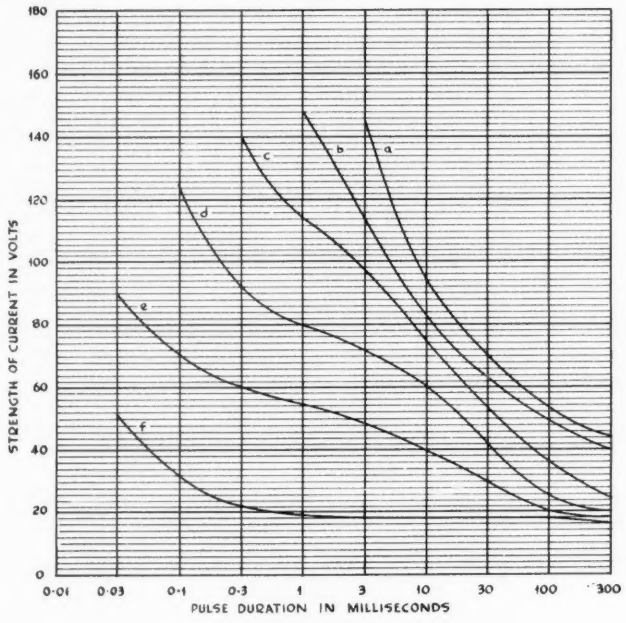


FIG. 2

Strength duration curves on the abductor pollicis brevis in a patient with a complete traumatic median nerve lesion at the wrist. (a) Twenty-one weeks after nerve suture the curve shows complete denervation. Curves (b) to (f) are taken at three-weekly intervals and show the progressive changes of reinnervation. Note the kinking and the shift of the curve to the left and its diminishing steepness.

The earliest sign of reinnervation in a previously completely denervated muscle on the SD curve is the appearance of a small discontinuity or kink in a curve that has hitherto been the smooth steeply rising curve of total denervation. This kink appears between six and eight weeks before there is clinical evidence of recovery. As regeneration proceeds, with more muscle fibres becoming reinnervated and the nerve component becoming more obvious, three changes are seen in the curve: the kink becomes wider, the curve shifts to the left, and its slope becomes less steep. Figure 2 shows the serial SD curves of a median nerve lesion that made steady progress to full reinnervation.

Once reinnervation has begun, the tests should be done every two weeks as this usually allows sufficient time to pass for a change in the curve to be seen. If the curve of a nerve lesion shows no further change over a period of six weeks it may be concluded that reinnervation

has ceased, and the reason must be sought. Figure 3 shows the curves in a patient with an ulnar nerve lesion who showed progress up to a stage where the muscle was graded 3—, but successive curves at two-week intervals after this showed no change. A neuroma was diagnosed and confirmed at operation.

Similar changes are seen in reverse in a degenerating lesion and again the early signs of denervation on the SD curve may precede clinical evidence of weakness and sensory loss by many weeks. The SD curve in the first two to three days after denervation is the double curve of partial denervation. The curve of complete denervation is always found within fourteen days of denervation. Figure 4 shows the changes in the SD curve in a patient with symptoms suggestive of ulnar neuritis without clear objective signs of sensory loss or muscle weakness.

Thus the plotting of a strength duration curve allows the detection of denervation, a fair

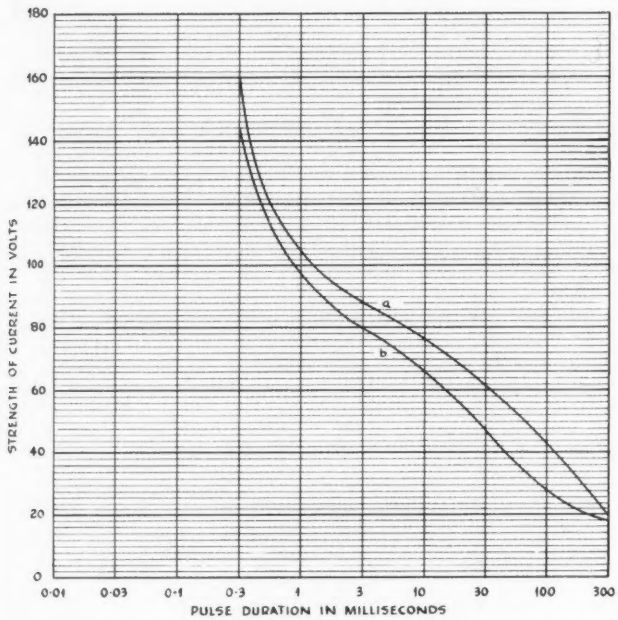


FIG. 3

Strength duration curves on the abductor digiti minimi in a patient with a traumatic lesion of the ulnar nerve at the level of the wrist, (a) twenty-four weeks after nerve suture showing the curve of partial innervation, and (b) five weeks later showing no change.

approximation of its extent, the detection of the early signs of reinnervation and the assessment of its progress.

The SD curve is the single most useful electrodiagnostic aid for the orthopaedic surgeon. Serial curves will afford reliable evidence of progress after nerve suture or of its arrest. They will afford early objective evidence of denervation in pressure syndromes of peripheral nerve injuries. They will allow the surgeon to decide whether a brachial plexus lesion is partial or complete at an early stage when pain, swelling and stiffness do not allow a detailed and reliable clinical assessment to be made. Plotting a curve on one muscle supplied by each root will give an accurate picture as to the extent and degree of involvement. The presence of a normal curve seven days after injury in a clinically complete nerve lesion strongly suggests a neurapraxia and therefore a good prognosis. Clearly the detection of a normal curve in a suspected hysterical palsy is extremely useful information.

It cannot be too strongly emphasised that provided the technique is accurate, the findings of an SD curve are very reliable. Modern electronic apparatus is thoroughly dependable and the wave forms vary less than 5 per cent. Thus detection of minor changes in a curve are significant.

Rheobase—It is claimed by some workers that the absolute value of the rheobase is significant. The value is said to be lower than normal in denervation, the denervated muscle being more excitable, and it is claimed also that a sudden rise in rheobase is significant of reinnervation. This is not true in the author's experience of over 300 patients with nerve lesions followed with SD curves plotted at intervals of two weeks from complete denervation to recovery (Wynn Parry 1954). Values may be normal throughout denervation, and in some cases may be actually higher.

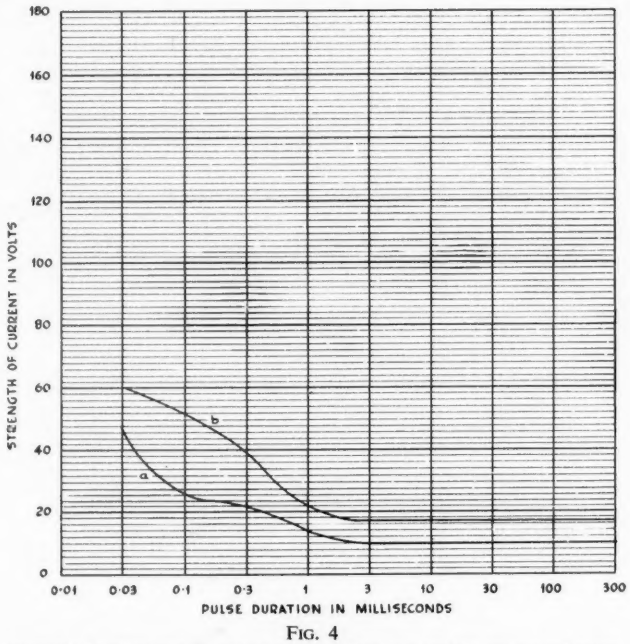


FIG. 4

Strength duration curves on the abductor digiti minimi in a patient suffering from an ulnar neuritis caused by compression at the elbow by callus, showing (a) slight evidence of denervation, and three weeks later (b) that there has been progressive denervation.

It needs to be stressed that the absolute values of an SD curve are not in themselves of significance. It is the shape and slope of the curve and any kinks that are of importance.

Chronaxie—This is a measurement of excitability of a tissue and was once very popular. It is defined as the time required for a current, of value twice the rheobase, to pass for the smallest contraction to be seen. Short values are indicative of nerve conduction, long values (3 milliseconds or more) of muscle stimulation. Unfortunately, intermediate values are not found and the chronaxie is more often than not still long in the presence of obvious reinnervation and short in the presence of clear signs of degeneration on the SD curve (Fig. 5). Chronaxie is therefore no longer used as an index of recovery.

Some workers use an abbreviation of the SD curve to save time. They measure the ratio of the smallest current to cause contraction at 1 millisecond to that at 100 milliseconds and express the result as the 100/1 millisecond ratio. Values of 2 or more are said to suggest

denervation, a value of 1 means normally innervated muscle, and values between 1 and 2, a partial lesion. This measurement suffers from exactly the same drawbacks as chronaxie, and values of 100/1 millisecond ratio of well over 2 may be found in muscles in which the SD curve shows an obvious kink.

As the full curve takes so little time to obtain and in view of the fallacies of abbreviations, it is strongly recommended that a full curve be plotted in all circumstances.

Nerve conduction—Before the SD curve is plotted it is important to carry out a test of nerve conduction. The nerve under study is stimulated by a short duration (1 millisecond or 0·3 millisecond) impulse using an active electrode over the nerve at its most accessible point and a dispersive electrode on the back of the neck (for nerves in the upper limb) or back of the

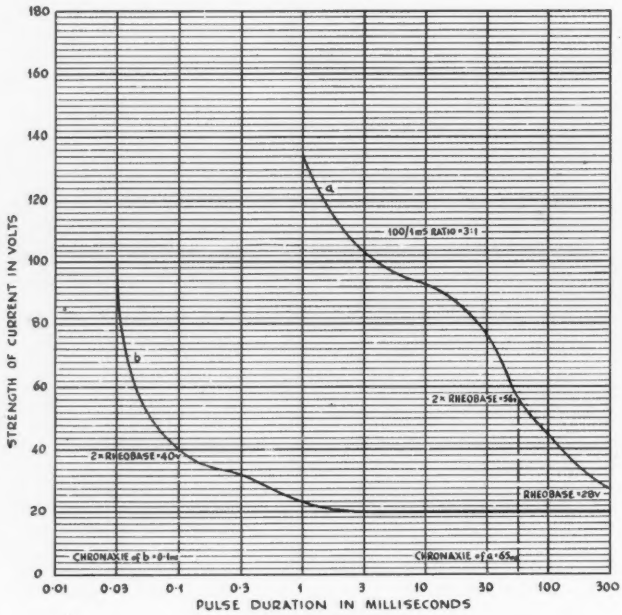


FIG. 5

(a) This strength duration curve on the abductor digiti minimi in a patient with a recovering ulnar nerve lesion after suture of the nerve shows that the chronaxie is long and the 100/1 milliseconds ratio is high, despite the obvious evidence of reinnervation. (b) is a strength duration curve on the abductor digiti minimi in a patient with an early ulnar neuritis. The chronaxie is short despite the obvious evidence of denervation.

lumbar spine (for nerves in the lower limb). The amount of current required to cause the slightest contraction in the muscles supplied by the nerve is measured and compared with that on the unaffected side.

Testing the nerve conduction is useful in three conditions. First, it enables one to find the level of the block in neurapraxia by stimulating the nerve at different levels in the limb. Secondly, it allows one to study the pattern of innervation and detect anomalies. This is particularly important in the hand, as one in five individuals has some anomaly of innervation and it is obviously important to find this out in suspected partial lesions. Thirdly, it allows a rapid survey of the extent of involvement in multiple lesions or root lesions—particularly in the arm.

The fifth and sixth cervical roots may be stimulated in the anterior triangle of the neck. The median, ulnar and radial nerves may be stimulated in the axilla. The median and ulnar

nerves can be stimulated at the elbow and wrist and the radial nerve in the middle of the upper arm. The femoral nerve may be stimulated in the femoral triangle, the sciatic nerve in the thigh, the medial popliteal nerve in the popliteal fossa and the lateral popliteal nerve behind the neck of the fibula.

An increase in the amount of current applied to the nerve that is necessary to produce contraction in the appropriate muscle is a sensitive index of early degeneration.

ELECTROMYOGRAPHY

The electromyograph machine is used to detect, amplify and display on a cathode ray screen the electrical activity associated with muscular contraction.

The motor unit consists of an anterior horn cell, its peripheral axon and a variable number of muscle fibres. The electrical activity connected with the smallest degree of voluntary movement is the discharge of a single motor unit. A needle electrode inserted into the muscle records this activity associated with the depolarisation of the muscle fibres of the unit as a

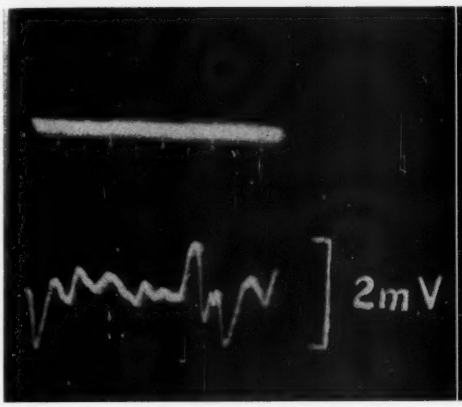


FIG. 6

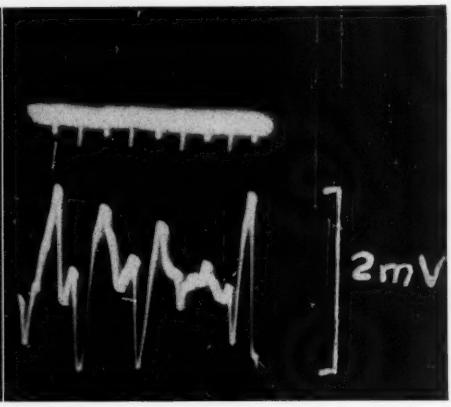


FIG. 7

Figure 6—Partial interference pattern of normal motor unit action potentials. Figure 7—Complete interference pattern of normal motor unit action potentials.

smooth diphasic wave with a duration of 5–7 milliseconds and an amplitude of 0·5–2 millivolts, which is called the motor unit action potential.

As voluntary contraction increases in power, more motor units come into action and their frequency of discharge increases, so that action potentials interfere with each other on the screen producing the so-called partial interference pattern (Fig. 6). On maximum volition so many units are discharging that the whole trace on the screen is distorted by motor unit action potentials—the so-called complete interference pattern (Fig. 7). Thus an idea of the number of motor units active on maximum volition can be gained by the appearance of the wave pattern.

Some ten to fourteen days after a muscle becomes denervated the individual muscle fibres begin to contract rhythmically and spontaneously. It is known that in the embryo muscle fibres contract spontaneously before they are innervated, and this fibrillation activity appears to be an inherent property of the muscle fibre. Denervation removes some trophic influence which normally prevents muscle from fibrillating.

Fibrillation activity is detected by noting spontaneous rhythmic spike potentials with a duration of 1 millisecond and an amplitude of 50–200 microvolts, giving a very characteristic high-pitched clicking noise in the loudspeaker (Fig. 8). Fibrillation persists until either the

muscle becomes reinnervated or becomes fibrotic, and may be found for many years after a peripheral nerve lesion that has not recovered. Detection of fibrillation is absolute evidence that some muscle fibres are denervated, and if no motor unit potentials can be found on attempted voluntary movement the lesion is complete. Sometimes fibrillation is not detected either because the muscle is cold or because the needle is not in the denervated area of a partially denervated muscle. The detection of fibrillation indicates denervation of the muscle fibres near the needle, and its absence does not necessarily mean that there is no denervation.

It will be realised that the fibrillation potential is the activity of one muscle fibre but that the motor unit action potential is the algebraic summation of all the fibrillation potentials in the motor unit under voluntary control. As the fibres are separated spatially the duration of the motor unit action potential is longer (5–7 milliseconds) than the fibrillation potential.

During regeneration of a peripheral nerve, action potentials on volition are seen which are highly polyphasic in shape (Fig. 9). Instead of the normal smooth diphasic action potential, the polyphasic potential is ragged, contains many spikes, and may be longer in duration than normal.

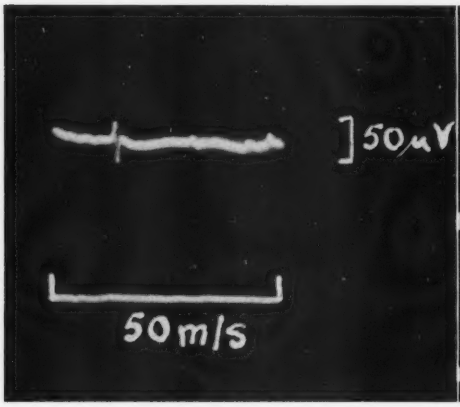


FIG. 8

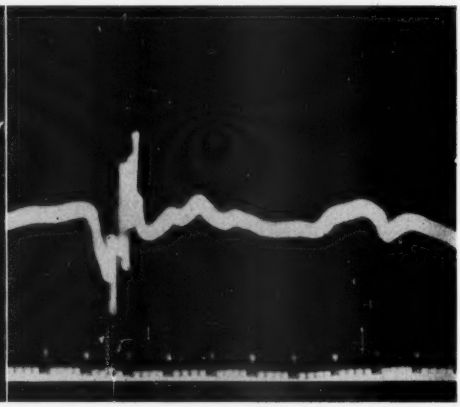


FIG. 9

Figure 8—Fibrillation potential indicating denervation. Figure 9—Polyphasic action potential of amplitude 1 millivolt recorded in a patient with a regenerating nerve lesion.

The explanation of this is that in the early stages of regeneration the myelin sheaths of the preterminal axons are not fully matured and thus the conduction velocity is slower than normal. This leads to temporal dispersion of the wave form and, as the conduction velocity varies in different nerve fibres within the unit, its form is ragged.

As regeneration proceeds the action potentials, which may start as short duration (2–3 milliseconds) polyphasic potentials, become bigger in amplitude and longer in duration, and, as more muscle fibres become reinnervated, more units come into action producing at first a discrete activity on extreme volition from a few units, later a partial interference pattern and finally a complete interference pattern. The same potentials are seen in reverse in degeneration of the peripheral nerve whether from trauma or from toxic, infective or metabolic peripheral neuritis. Thus the E.M.G. picture of a polyneuritis is of polyphasic action potentials, becoming shorter in duration and the interference pattern becoming progressively reduced. The conduction velocity, to be discussed in detail later, becomes much slower than normal in peripheral neuropathies.

In chronic, slowly degenerating lesions, the surviving intact axons send out collateral sprouts from their endings to reinnervate the denervated fibres. It seems that the denervated fibres produce a neurotropic substance, identified as an unsaturated fatty acid, which attracts

these sprouts. Thus the denervated fibres become " mopped up " as it were, by intact axons. The result on the E.M.G. is the detection of giant motor units, for these units contain many more muscle fibres than normal, thus giving rise to a motor unit of long duration (15 milliseconds or more) and high amplitude (up to 10 millivolts). Moreover these units come into action on the slightest voluntary activity. Large units appear in a normal muscle occasionally but only on the greatest exertion and they are never as large as these giant pathological units; it is the appearance of the latter that is so typical of a chronic progressive lesion of the lower motor neurone.

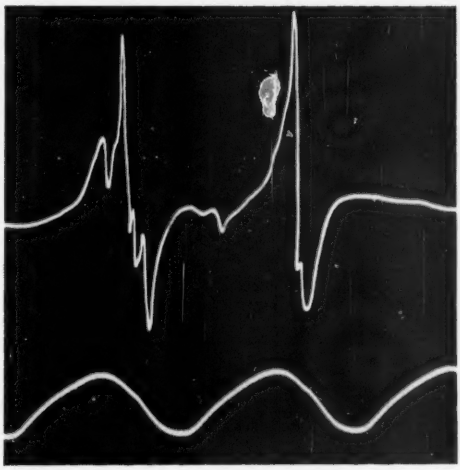


FIG. 10
Giant motor unit action potential of amplitude 6 millivolts and duration 12 milliseconds, seen in a partial median nerve lesion.

Because the majority of slowly progressive degenerative lesions of the lower motor neurone are spinal cord diseases affecting the anterior horn cells—such as syringomyelia and motor neurone disease—it has been assumed that the characteristic picture of these lesions is the detection of giant units indicating voluntary contraction, but with a greatly reduced interference pattern from the loss of many units. Although this assumption is almost always reliable in practice, it must be stressed that such a picture merely indicates a chronic lesion, which is usually due to spinal cord disease. For instance, the same appearance may be seen in the abductor pollicis brevis with a partial traumatic median nerve lesion at the wrist (Fig. 10) which would not, of course, give rise to difficulty in diagnosis. In a chronic polyneuritis, giant potentials may be seen, but polyphasics and only a slightly reduced interference pattern are the concomitant features that usually clarify the picture.

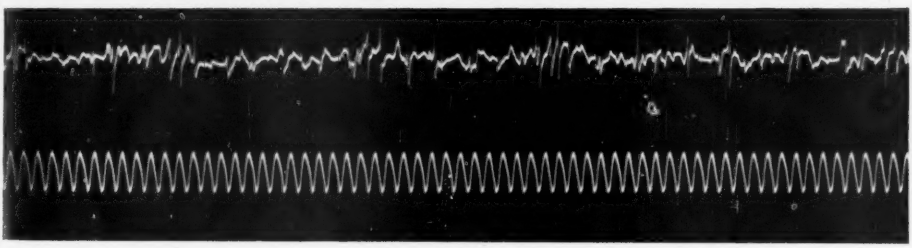


FIG. 11
The action potentials in myopathy on volition have an extremely short duration and low amplitude. There are 2 milliseconds between the peaks on the time scale.

When a disease process such as a myopathy affects the muscle fibre itself, the motor unit potentials seen on the E.M.G. on volition become much shorter and polyphasic because individual muscle fibres within the unit have been lost. But because whole motor units have not been lost there is a normal interference pattern until a late stage. The combination of a full interference pattern of motor unit activity with very short duration polyphasic action potentials is most characteristic of a myopathy (Fig. 11).

In recent years a group of disorders has been recognised, and called as a whole neuromyositis, in which the common feature is an abnormality both of the muscle fibre and

the terminal part of the axon. It is known that the neuromuscular junction is a highly vulnerable part of the body and, as such, is liable to be readily affected by disease. The exact nature of the disorder is unknown, though the evidence favours a metabolic abnormality. The clinical features are those of wasting and weakness of the proximal limb girdle muscles, so that patients complain of difficulty in walking up stairs and in reaching above shoulder level. There are four main diseases which give rise to this picture: 1) certain collagen disorders, notably systemic lupus erythematosus, dermatomyositis, and idiopathic polymyositis; 2) the carcinomatous neuromyopathies, usually associated with carcinoma of the bronchus, in which the patient may present with wasting and weakness of the girdle muscles many months or years before signs of the carcinoma become obvious; 3) thyrotoxic myopathy, and 4) the rare myopathy associated with myasthenia gravis. All these conditions have an E.M.G. picture with polyphasic action potentials on volition and a reduced interference pattern—indicating disorder of nerve (and there may be changes on the SD curve) and also areas where the action potentials are very short in duration and of very high frequency—indicating a myopathic lesion. These abnormalities are characteristically patchy and, on exploring with the needle, many areas are found to be normal, all of which is typical of a neuromyositis with a mixed myopathic and neuropathic lesion.

In the true heredo-familial myopathies no long duration polyphasics are seen, the interference pattern is full and all the muscle is uniformly affected, so usually there is no difficulty in distinguishing a true myopathy from a neuromyositis.

Conduction velocity—Measurement of motor and sensory conduction velocities can be of great value in diagnosis and localisation of lower motor neurone lesions.

The earliest electrical evidence of interference with nerve activity is a slowing of conduction velocity. In lesions produced by pressure it can be shown to occur some weeks before the SD curve or the E.M.G. show any abnormality.

The technique is as follows. A needle electrode is inserted into the appropriate muscle, and its nerve stimulated at a convenient point. The stimulus triggers the E.M.G. time base and is increased until the summated motor unit action potential is seen on the screen. A pulse duration of 0·1 millisecond is usually used and a supramaximal stimulus given so as to stimulate all the motor units available. The time from stimulus to start of action potential is now measured and expressed in milliseconds. Normal values have been established, though conduction velocity can of course be expressed in metres per second by measurement of distance between stimulus and the needle electrode. Figure 12 illustrates this.

This technique of measuring motor conduction time is of value in conditions such as the carpal tunnel syndrome and ulnar neuritis. For the former, a needle electrode is inserted into the abductor pollicis brevis and the median nerve stimulated first at the elbow, and then at the wrist, and the conduction time measured. If there is slowing of conduction due to pressure at the wrist, both values will be longer than normal, but by subtraction it will be found that the time from elbow to wrist is normal and that from wrist to the abductor pollicis brevis is prolonged. Normal values from elbow to short abductor are 8–12 milliseconds and from wrist to short abductor 4–6 milliseconds. Thus if the time from elbow to short abductor was 24 milliseconds and from wrist to short abductor 18 milliseconds, it is clear that the time from elbow to wrist is 6 milliseconds (normal) and the slowing is due to a lesion at the level of the wrist (Fig. 13).

Similarly, if there are symptoms of ulnar neuritis, it is easy to establish if the slowing of conduction is occurring at the level of the wrist or at the elbow. If the slowing is uniform down the nerve it strongly suggests that the lesion responsible is in the nerve roots.

In a recovering peripheral nerve lesion motor conduction velocity is much slower than normal, but as recovery proceeds so the conduction velocity increases. Conversely, degenerating lesions of the peripheral nerves—whether traumatic, toxic, metabolic or infective—have a slowing down of conduction velocity.

In spinal cord lesions the intact axons conduct at a normal rate and this is a valuable distinguishing sign between an anterior horn cell lesion and a lesion of the peripheral axons.

In a peripheral nerve disorder not only may the conduction time be prolonged, but the action potential resulting from supramaximal nerve stimulation may well be much longer in duration than normal, or " dispersed " as it is called. Thus the action potential in response to stimulation may be 50 milliseconds in duration and highly polyphasic in the carpal tunnel syndrome because of the different degrees of slowness of conduction in the axons compared with the normal smooth diphasic response of 10–18 milliseconds duration.

Sometimes this dispersion may be the only abnormality because some fibres are conducting normally; but inspection of the duration and nature of the action potential will show if the

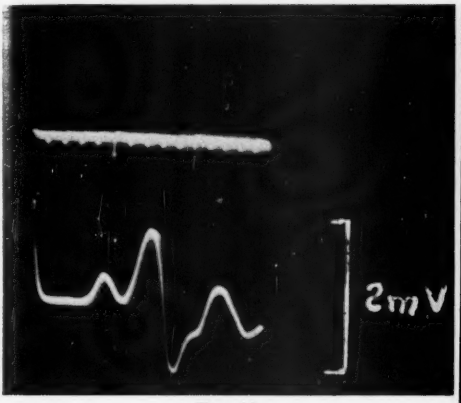


FIG. 12

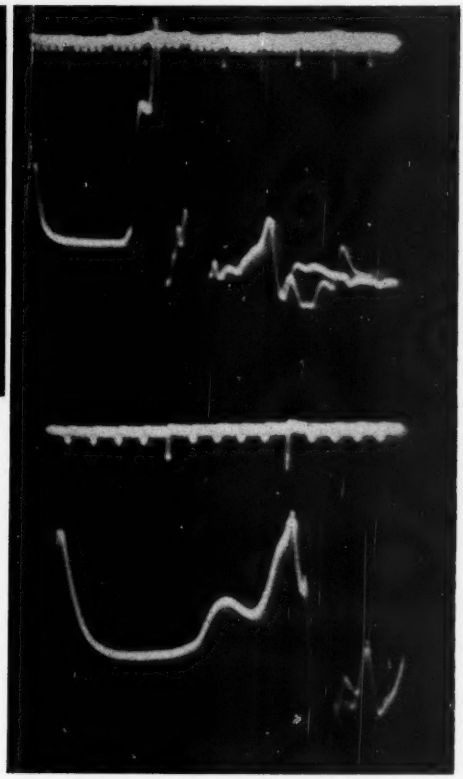


FIG. 13

FIGS. 12 AND 13

Figure 12—An electromyograph of the motor conduction time in normal muscle, in which the stimulus artefact is on the extreme left of the trace. There is then a delay of 4 milliseconds before the appearance of the motor unit action potential. This recording was made with the stimulus over the median nerve at the wrist and the needle electrode in the abductor pollicis brevis. Figure 13—Motor conduction tests in a patient with a suspected carpal tunnel syndrome. The needle electrode is in the abductor pollicis brevis; the stimulus is at the elbow in the upper trace, and at the wrist in the lower trace. The conduction time from the elbow to the thumb is 12 milliseconds and from the wrist to the thumb is 6 milliseconds. There is slight slowing at the level of the wrist but the obvious abnormality is the gross dispersion of the wave form which occupies some 30 milliseconds, which indicates extreme slowing of conduction in many fibres.

dispersion indicates slowing in many axons. Therefore it is important to look not only at the conduction time but the degree of dispersion of the action potential resulting from nerve stimulation.

Sensory conduction—In the last two years the technique of recording sensory nerve action potentials percutaneously in response to stimulation of the sensory nerve distally has been perfected (Gilliatt and Sears 1958). Two ring electrodes are applied to the finger—one at the base and one round the middle phalanx and connected to the stimulator of the E.M.G. machine which triggers the time base.

Silver surface electrodes are applied over the nerve at the wrist or elbow and connected to the E.M.G. machine. An earth is arranged between stimulating and pick-up electrodes.

The digital branch of the median nerve is stimulated in the index finger and the pick up electrodes arranged over the nerve at the wrist, while the digital branch of the ulnar nerve is stimulated on the little finger and the pick up electrodes arranged over the nerve at the wrist.

The sensory action potential is only 10–30 microvolts in amplitude so that a high gain amplifier must be used and perfect relaxation obtained, as interfering motor unit potentials

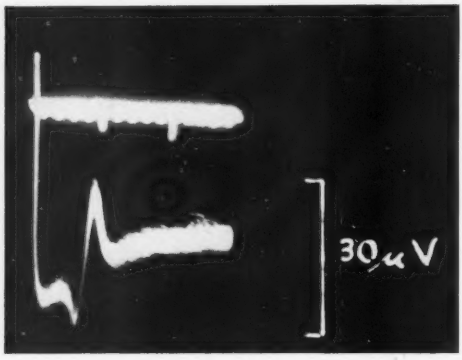


FIG. 14

This normal sensory nerve conduction trace was recorded by stimulating a digital nerve of the index finger with ring electrodes; the sensory action potential was recorded over the median nerve at the wrist. The stimulus artefact appears on the extreme left of the trace and the sensory action potential of 30 microvolts amplitude is seen 2 milliseconds later.

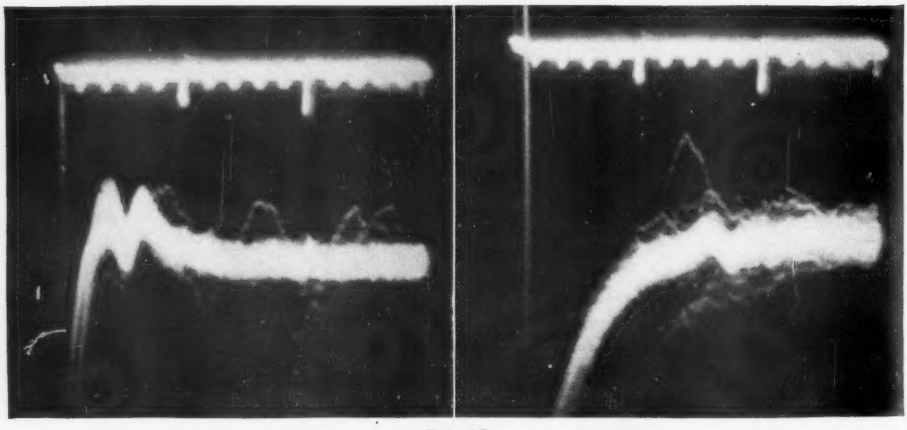


FIG. 15

The tracing on the left shows the normal pattern of median nerve sensory conduction recorded at the wrist after stimulation of the digital nerve of the index finger; and, on the right, the pattern in an early carpal tunnel syndrome. The conduction time is considerably prolonged (7 milliseconds instead of 2) and the amplitude is grossly diminished.

picked up by the electrodes are 50 to 100 times as great. However, with care, this technique is relatively easy and well worth while taking time and trouble to perfect.

The normal conduction time from finger to wrist is 2–4 milliseconds, and with pressure on the nerve due to the carpal tunnel syndrome, for example, this time may be prolonged to two or three times the normal, and the amplitude of the action potential is reduced to

microvolts or even disappears altogether. It is because absence of a sensory nerve potential is so significant that the technique must be good, for if no action potential is seen, either the sensory nerve is diseased or the technique is faulty and the action potential is not being recorded.

The median or ulnar nerve may be stimulated at the wrist and the action potential picked up at the elbow; but here the action potential is bigger, not only because the sensory action potential is picked up but also because the antidromic impulses are also travelling up the motor nerve, so that the amplitude may reach 50 microvolts. The measurement of these velocities allows the site of the nerve lesion to be determined. The extreme sensitivity and practical value of this test is shown by the fact that significant slowing of sensory conduction, with diminution in the amplitude of the action potential, can be seen in the carpal tunnel syndrome, when there is a normal motor conduction time, a normal SD curve and a normal E.M.G. (Figs. 14 and 15).

The detection of the sensory action potential in response to stimulation of the growing nerve end through the skin affords valuable evidence of satisfactory progress after nerve suture, particularly when many months may elapse before a muscle can be reinnervated, and the SD curve and E.M.G. show signs of recovery.

This is done by placing the pick up electrodes at an accessible part of the nerve proximal to the level of the lesion and while the stimulating electrode is moved up the expected path of the growing nerve, starting distally, until a sensory action potential is detected. This gives the level to which the nerve has grown. This test is particularly useful in ulnar nerve lesions between the wrist and the elbow.

CONCLUSIONS

Electrodiagnosis is of value to the orthopaedic surgeon in the diagnosis and prognosis of lower motor neurone lesions, and in the localisation of the site of the lesion, whether in the spinal cord, the peripheral axon, or the muscle. It must be stressed that the greatest value is obtained from these tests by completing the full electrodiagnostic procedure, which includes nerve conduction, the SD curve, the E.M.G., and, when indicated, the motor and sensory conduction times.

The various tests also offer sensitive evidence of regeneration and degeneration of a peripheral nerve some weeks before clinical evidence is manifest. They are of much help in making the decision whether to explore a nerve or plexus lesion or not. They can offer valuable evidence of satisfactory progress after nerve suture and can detect at an early stage any hold up in recovery.

They can offer helpful information as to the site of a lower motor neurone lesion and can distinguish a myopathic, neuropathic and spinal cord lesion, as in the differential diagnosis of wasting of the small muscles of the hand.

The techniques of plotting SD curves and nerve conduction are so simple, reliable and easily learnt that they deserve to be used routinely by those concerned with peripheral nerve surgery. The technique of electromyography is more complex and requires wide experience, but a knowledge of the information it can supply and the indications for its use should be of great help to the surgeon confronted with a difficult problem in the diagnosis or prognosis of a lower motor neurone lesion.

SUMMARY

1. The results of electrodiagnostic tests reveal the level and progress of nerve lesions.

2. The findings in a myopathic lesion are: *a*) a normal SD curve; *b*) no fibrillation on the E.M.G.; *c*) on volition a full interference pattern—because motor units have not been lost, but only muscle fibres within the units; *d*) the action potentials are short in duration, polyphasic and of low amplitude; *e*) conduction times are normal.

3. In peripheral nerve affections these findings are evidence of denervation: *a*) a sluggish response to a long duration current; *b*) a kinked SD curve; *c*) fibrillation at rest on the E.M.G.;

d) a reduced interference pattern of motor unit action potentials on volition; *e*) a preponderance of polyphasic action potentials of long and short duration; *f*) slowing of conduction times.

4. Evidence of reinnervation is given by: *a*) a kink in the SD curve; *b*) the appearance of a response in the muscle to nerve stimulation; *c*) a progressive increase in conduction velocity; *d*) the appearance of polyphasic action potentials on volition; *e*) fibrillation becoming more difficult to detect.

5. The following findings suggest a lesion of the anterior horn cell: *a*) difficulty in detecting fibrillation, and a relatively normal SD curve in the presence of marked wasting. (Intact axons have "mopped up" denervated fibres.) *b*) Marked reduction in interference pattern with 'giant units'. *c*) Normal conduction velocity.

I wish to thank the Director General of Medical Services, Royal Air Force, for permission to publish this paper.

REFERENCES

ERB, W. (1883): *Handbook of Electrotherapeutics*. Translated by L. Putzel. New York: William Wood & Co.

GILLIATT, R. W., and SEARS, T. A. (1958): Sensory Nerve Action Potentials in Patients with Peripheral Nerve Lesions. *Journal of Neurology, Neurosurgery, and Psychiatry*, **21**, 109.

RITCHIE, A. E. (1954): Electrical Diagnosis. In *Medical Research Council Special Report Series* No. 282 *Peripheral Nerve Injuries*, pp. 239–262. London: Her Majesty's Stationery Office.

WYNN PARRY, C. B. (1954): Electrical Methods in Diagnosis and Prognosis of Peripheral Nerve Injuries and Poliomyelitis. *Brain*, **76**, 229.

THE BROKEN SCAPHOID BONE

The Case Against Pessimism

P. S. LONDON, BIRMINGHAM, ENGLAND

From the Birmingham Accident Hospital

The management of fractures of the carpal scaphoid bone is based on two premises: firstly that failure of bony union is common and secondly that failure of union causes serious disability. It is generally agreed that successful treatment depends on prompt immobilisation continued without remission until union has occurred, but opinions have varied as to how to immobilise the fracture.

IMMOBILISATION

The wrist—At one time or another most of the possible positions of the wrist have been recommended. Snodgrass (1933) used a flat palmar splint. Speed, quoted by Berlin (1929), advised slight flexion and Hosford (1931) hyperextended the wrist. Others (Berlin 1929, Soto-Hall and Haldeman 1934, Friedenberg 1949) advocated extension and radial deviation because dissections led them to believe that this reduced and impacted the fracture. These recommendations, however, did not take account of the fact that dissection and division of the bone is unlikely to reproduce exactly the conditions of natural fractures, and McLaughlin (1954) found during operations on fractured scaphoid bones that no one position of the wrist could be relied upon either to reduce or to immobilise the fracture. He stressed the importance of stability. The usual practice now is to immobilise the wrist in slight extension and ulnar deviation (Watson-Jones 1955, Bunnell 1956).

The thumb—Soto-Hall (1945) advised that the thumb be fixed in abduction and extension because dissection and experiment led him to believe that this prevented movements that could disturb the fracture. It is more usual, however, to fix the thumb in a useful position, roughly in line with the radius (Watson-Jones 1955, Key and Conwell 1956, Adams 1957). Against the prevailing view that the thumb should be encased is Böhler's experience (Jahna 1954, 1955) that it can safely be left free.

Extent of immobilisation—Extensive immobilisation such as encasing the elbow (Grace 1929) or the fingers (Nenninger 1955) has not apparently been justified by the results. Indeed, Hicks (1956) has shown that instead of preventing disturbance of a fracture by muscular activity, a well fitting plaster may in some instances actually facilitate it by immobilising the joints more effectively than the fracture. Nevertheless, Squire (1959) has made a good case for enclosing the forearm and elbow with the forearm fully supinated in the uncommon case with pronation (flexion) deformity at the site of fracture.

Period of immobilisation—Although there has been disagreement about how to immobilise the fracture, it is generally agreed that prolonged immobilisation may be necessary, especially for the older fractures and fractures in the proximal third of the bone (Watson-Jones 1934, 1955; Dickison and Shannon 1944; Stewart 1954). Jahna (1955) applied to the scaphoid bone Pauwels's (1935) views on fractures of the neck of the femur and recommended that the closer the fracture line is to the long axis of the limb, the longer the period of immobilisation should be. The results obtained by these workers (Table I) show that for fresh fractures near the middle of the bone an average time of two to three months in plaster enabled nearly all to unite, while for older and proximal fractures much longer periods of immobilisation were employed. Six to seven months in plaster has not been regarded as remarkable and in one case Robertson and Wilkins (1944) persisted with plaster for sixty weeks before diagnosing union. They commended the man's patience but there seems to have been little else to commend in this instance.

The exact incidence of union by bone is not certain because not all patients complete treatment, but the data reproduced in Table I suggest that about 95 per cent of all fractures under a month old unite with proper treatment. This is in contrast with other published opinions but accords with our own experience.

THE AUTHOR'S PRACTICE

For some years the policy at the Birmingham Accident Hospital has been at variance with that of insistence on immobilisation until either bony union is complete or non-union established. The emphasis has been on treating the wrist rather than its radiological appearance. The wrist and thumb are immobilised in a useful position for six to eight weeks, the plaster being repaired or replaced as required. At the end of this time, provided the patient has a comfortable wrist, a reasonable range of movement and a good grip with little or no local tenderness, the plaster is usually discarded irrespective of the radiological appearances. If, on the other hand, at this time there is discomfort, much weakness or pain or tenderness, the plaster is reapplied and kept on for a further period of four to six weeks. After about three months in plaster such a wrist is usually left free and helped to regain its function. Sometimes plaster is followed for a week or two by strapping or occasionally by a leather shield.

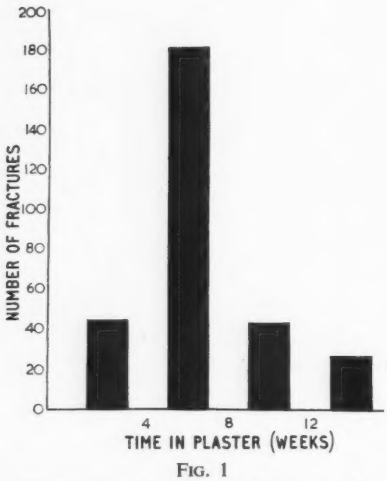


FIG. 1

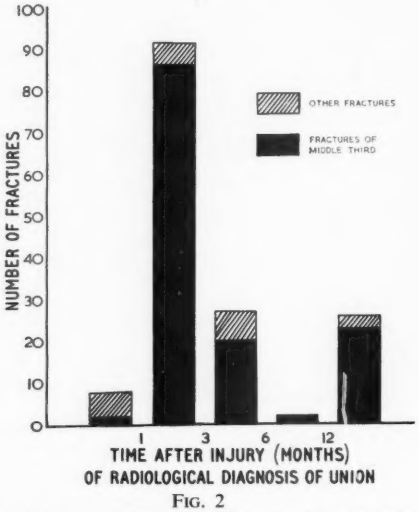


FIG. 2

Figure 1—Time in plaster: 295 fractures of the scaphoid bone. (In five of the 300 cases the period of immobilisation was not known.) Figure 2—Time to radiological diagnosis of union.

RESULTS

Two-thirds of 300 fresh fractures were in plaster for eight weeks or less and only one in ten for more than three months (Fig. 1). In some fifty cases there was no union radiologically* for at least three months after fracture (Fig. 2). In some cases (Fig. 2) twelve months or more elapsed before union took place and this occurred even though the wrist was in active use without any form of protection for most of this time. An illustrative case is shown in Figures

* Radiological union: this implied normal texture throughout the bone in antero-posterior, lateral and two oblique views. In other cases union was diagnosed because although the last available radiological appearances did not quite reach this unequivocal standard, they had improved steadily and the wrist was strong, supple and painless.

3 to 6. It is also noteworthy that in about fifty cases union occurred in spite of delay in treatment, inadequate immobilisation and other adverse factors.

Bony union occurred in about 95 per cent of the 227 out of the 300 fresh fractures which could be studied adequately (Fig. 7). The findings are similar to those recorded in Table I, and, as in two of the series recorded there, many patients were lost to sight before the degree of union could be determined.

FIG. 3

FIG. 4

FIG. 5

FIGS. 3 TO 6

Illustrating union occurring unexpectedly. The case of a man aged twenty-four who broke his right wrist. Figure 3—Appearance on day of injury, January 1954. Plaster immobilisation begun then and continued for fourteen weeks. Figure 4—Nine months after injury. Appearances strongly suggest non-union. Figure 5—Sixteen months after injury. Possibly uniting after all. Figure 6—Twenty-six months after injury. United.

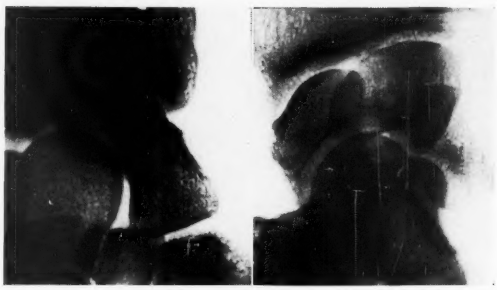


FIG. 6

DISCUSSION

If the figures given in Table I and Figure 7 be accepted and if we accept that the untraced fractures did well, one may ask why the fracture has such a bad reputation. The explanation seems to lie in what has been written about this fracture. Until Röntgen discovered x-rays in 1895 fracture of the scaphoid bone was known only as an occasional finding at necropsy (Callender 1866). Within five years of its becoming possible to demonstrate the fracture during life Stimson (1902) stated that " total disability of the wrist " was the normal result of this injury. A generation later Adams and Leonard (1928) wrote " it is an undisputed fact, however, that in the vast majority of cases the bone does not unite." In 1957 Adams placed the scaphoid bone second only to the neck of the femur in its liability to non-union and recently Decoulx, Razemon and Lemerle (1959) reported non-union in 36 per cent of a series treated in plaster. The reason for the early pessimism is suggested in a paper by Codman and Chase (1905). They recommended a month in splints, a month of massage and then operation if the fracture had not united. In fact, some of their patients appear to have worn splints for only a week or so. It seems clear, therefore, that the bad reputation was based originally upon inadequate treatment and an impatient resort to operation combined with failure to distinguish between the effects of the fracture and the effects of the treatment inflicted on the wrist. This reputation has evidently been maintained by numerous reports of

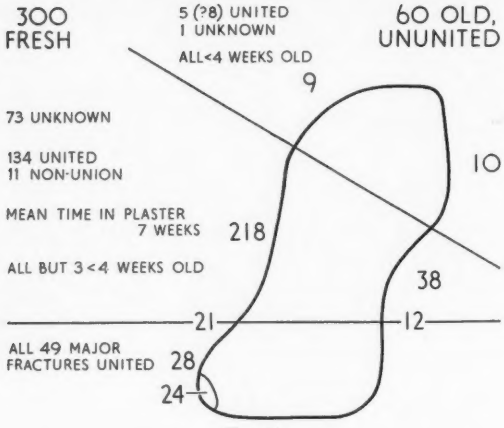
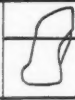
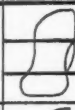
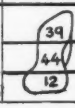


FIG. 7

The Birmingham Hospital series: summary of the results of treatment of 300 fresh fractures. Sites of injury in sixty ununited fractures presenting with symptoms in the wrist. The sixty ununited fractures are entirely separate from the 300 fresh ones.

TABLE I

RESULTS OF TREATMENT OF FRACTURE OF THE SCAPHOID BONE. FOUR LARGE SERIES

Source	Time interval injury/ treatment (*weeks*)	Site of fracture	Average time in plaster (*weeks*)	Number of fractures	Union	Remarks
Watson-Jones (1934)	Less than 4		22	6	5	
			8	66	66	
	4 to 52		30	23	22	
Dickison and Shannon (1944)	Less than 1		20	12	8	
			12	118	112	Four out of six displaced fractures did not unite
	1 to 5		12	3	2	
			12	12	12	Middle third only
Stewart (1954)	Less than 4		23 12 8	323	258	62 results unknown 3 non-union
	Over 4		20	99	51	36 results unknown 3 non-union
Jahna (1954) (1955)	" Recent fractures "		7–9	734	557	154 unknown 23 non-union
	" Older fractures "		10–20 11–26 13	116	95	Different periods in plaster according to obliquity of fracture

disabling effects of non-union. Non-union that has not required treatment, on the other hand, has only rarely been mentioned in the literature (Perkins 1950, Scott 1956).

Non-union—Sixty patients with established non-union seen at this hospital* have done much to dispel gloom about the condition. It was striking to find that all but seven of these patients came to the hospital because their wrists had recently become painful after an injury or strenuous use (Table II). Many had previously had little or no trouble from their wrists (Fig. 8) and some insistently denied having had any, in spite of vigorous use. Forty-nine of these sixty patients were able to return to work after two to four weeks in plaster or strapping but four of them had to be provided with a leather shield. Eleven were operated on. The operation was not always a formal arthrodesis of the wrist. In some instances it was sufficient for small spurs of bone to be removed. Partial or complete excision of the scaphoid bone has not been favoured in this hospital, and though numerous cases have been reported it is hard to find this type of operation recommended without qualification.

FIG. 8

Non-union of fracture of scaphoid bone of over thirty years' standing, detected when pain led to radiological examination. The patient, a man aged forty-nine, had suffered a painful right wrist in boyhood but had had no treatment. Pain had subsided and the wrist had given no trouble during the intervening years.

These observations suggest strongly that non-union is not itself a cause of symptoms but that it predisposes a wrist to react painfully to injury or overuse. Strong support for this suggestion is provided by comparing the distribution of fresh and ununited fractures between

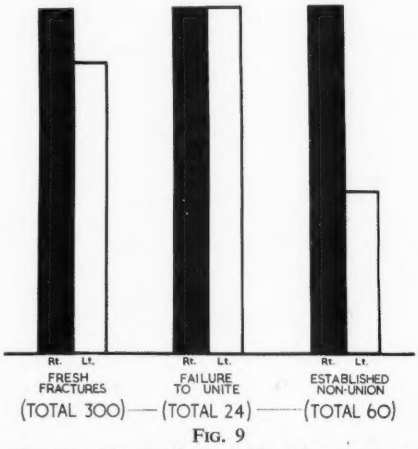


FIG. 9

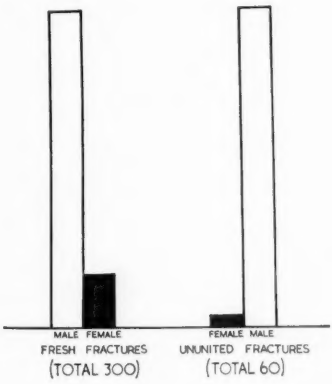


FIG. 10

Figure 9—The distribution of fresh fractures between the two wrists. Failure of fresh fractures to unite. Established non-union presenting with symptoms. Figure 10—Distribution of fresh and ununited fractures between the sexes.

he wrists (Fig. 9) and between the sexes (Fig. 10). Whereas the two scaphoid bones are bout equally liable to fresh fractures and to failure of bony union, non-union was seen wice as often in the right wrist as in the left. This makes it reasonable to suppose that right

* These form a series separate from the 300 fresh fractures.

TABLE II

CIRCUMSTANCES OF DIAGNOSIS OF NON-UNION OF FRACTURE OF SCAPHOID BONE

Radiograph taken because of:	Number of wrists
Pain after injury . . .	48
Pain after use . . .	5
Miscellaneous reasons: lump, 1; swelling, 1; sudden pain, 1; injury other wrist, 1	4
No reason obvious . .	3
Total . . .	60

TABLE III

AGE OF FRACTURE AND PRESENCE OF OSTEOARTHRITIS

Age of fracture	Number of wrists		Osteoarthritis (number of wrists)
Less than 5 years	19	24	12
6 to 10 years .	5		
11 to 20 years .	3	11	7
Over 20 years .	8		
Unknown. .	25		17

wrists with ununited scaphoid bones are more vulnerable—and more liable to give trouble—than left wrists, which are less used by most people and less liable to injury. Though the number of women is too small for the figures to be significant, the low incidence in women of established non-union with symptoms is consistent with the idea of fresh injury as the provocative cause and non-union as the predisposing cause of symptoms.

Osteoarthritis—Osteoarthritis is commonly regarded as the disabling consequence of non-union but it is neither inevitable nor, when present, necessarily a cause of symptoms (Scott 1956). In general, the older the fracture the greater the likelihood of osteoarthritis (Table III), but even after thirty years there may be none (Fig. 11), and some badly degenerate wrists (Fig. 12) are so free from symptoms as to be regarded as almost normal.

FIG. 11

FIG. 12

Figure 11—Non-union of thirty years' duration in the right wrist of a man aged fifty-one. No arthritis in spite of degenerate proximal fragment. Figure 12—Severe open fracture-dislocation of left carpus with loss of proximal fragment of scaphoid bone in a man aged forty-five. In spite of its disorganised appearance this wrist was not completely stiff and stood up to manual labouring.

CONCLUSIONS

Acceptance of these ideas will mean a considerable saving in working time and waiting time for patients, and lessened demand on plaster rooms and fracture clinics, for the need to continue immobilisation beyond eight to twelve weeks is questionable. If the wrist has become strong and comfortable the patient will suffer from the inconvenience of the plaster with doubtful benefit, for there is no conclusive evidence that prolonged immobilisation does in fact prevent non-union. Bony union can occur without it; fibrous union can occur in spite

of it. Even though the fracture line is still visible when the plaster is discarded it may nevertheless be obliterated during the ensuing six to twelve months. Even when fibrous union does develop it is not usually disabling; it is most often brought to light only by some injury and the consequent symptoms usually subside within a month. Delayed treatment, fractures in the proximal third and disturbed circulation of the bone are unfavourable factors, but the fate of a fracture cannot be foretold with accuracy during the first few weeks. The initial radiographs of twenty-four* fractures that were seen to progress from the fresh state to established non-union could be matched in all other respects by those of twenty-four fractures that united by bone. Even if the prospects can be foreseen there is no certain way of achieving union.

It seems that a plan of treatment based on symptoms rather than on radiological signs is justified by the results. Operation should be reserved for cases in which symptoms persist and disable in spite of suitable supports.

SUMMARY

1. The bad reputation of fractures of the scaphoid bone is based on *a*) past disasters caused by inadequate splintage and premature resort to operation; and *b*) selection of disabling cases for publication.

2. The results obtained in five series comprising over 1,000 fractures suggest that 95 per cent of adequately studied fractures less than a month old unite if properly treated.

3. Sixty cases of established non-union have provided evidence that disability is almost always the result of further injury and that the disability is usually relieved by a short time in plaster.

4. There is no evidence to support the widely held beliefs that *a*) union by bone can occur only if the fracture is immobilised throughout the process; and *b*) union after prolonged immobilisation is due to the immobilisation.

5. A policy of treating the wrist and not merely its radiological appearances is advocated.

I am much indebted to my colleagues for their co-operation and to the patients who attended for re-examination.

REFERENCES

ADAMS, J. C. (1957): *An Outline of Fractures.* Edinburgh and London: E. & S. Livingstone Ltd.

ADAMS, J. D., and LEONARD, R. D. (1928): Fracture of the Carpal Scaphoid. *New England Journal of Medicine*, **198**, 401.

BERLIN, D. (1929): Position in the Treatment of Fracture of the Carpal Scaphoid. *New England Journal of Medicine*, **201**, 574.

BUNNELL, S. (1956): *Surgery of the Hand.* Third edition. Philadelphia and London: J. B. Lippincott Company.

CALLENDER, G. W. (1866): Fracture of the Carpal End of the Radius, and of the Scaphoid Bone. *Transactions of the Pathological Society of London*, **17**, 221.

CODMAN, E. A., and CHASE, H. M. (1905): The Diagnosis and Treatment of Fracture of the Carpal Scaphoid and Dislocation of the Semilunar Bone. *Annals of Surgery*, **41**, 321.

DECOULX, P., RAZEMON, J. P., and LEMERLE, P. (1959): Les fractures et pseudarthroses du scaphoide carpien. *Lille Chirurgical*, **14**, 113.

DICKISON, J. C., and SHANNON, J. G. (1944): Fractures of the Carpal Scaphoid in the Canadian Army. *Surgery, Gynecology and Obstetrics*, **79**, 225.

FRIEDENBERG, Z. B. (1949): Anatomical Considerations in the Treatment of Carpal Navicular Fractures. *American Journal of Surgery*, **78**, 379.

GRACE, R. V. (1929): Fracture of the Carpal Scaphoid. *Annals of Surgery*, **89**, 752.

HICKS, J. H. (1956): *Personal communication.*

HOSFORD, J. P. (1931): Prognosis in Fractures of the Carpal Scaphoid. *Proceedings of the Royal Society of Medicine* (Section of Surgery), **24**, 982.

JAHNA, H. (1954): Behandlung und Behandlungsergebnisse von 734 frischen einfachen Brüchen des Kahnbeinkörpers der Hand. *Wiener Medizinische Wochenschrift*, **104**, 1,023.

* Thirteen of these were separate from the main series of 300.

JAHNA (1955): Die konservative Behandlung des veralteten Kahnbeinbruchs der Hand. *Verhandlungen der Deutschen Orthopädischen Gesellschaft*, **43,** 156.

KEY, J. A., and CONWELL, H. E. (1956): *The Management of Fractures, Dislocations and Sprains.* Sixth edition. London: Henry Kimpton.

MCLAUGHLIN, H. L. (1954): Fracture of the Carpal Navicular (Scaphoid) Bone. Some Observations Based on Treatment by Open Reduction and Internal Fixation. *Journal of Bone and Joint Surgery*, **36–A,** 765.

NENNINGER, W. (1955): Über die Behandlung von Kahnbeinfrakturen der Hand. *Deutsches Medizinisches Journal*, **6,** 224.

PAUWELS, F. (1935): *Der Schenkelhalsbruch. Ein mechanisches Problem.* Stuttgart: F. Enke.

PERKINS, G. (1950): Fractures of the Carpal Scaphoid. *British Medical Journal*, **i,** 536.

ROBERTSON, J. M., and WILKINS, R. D. (1944): Fracture of the Carpal Scaphoid. *British Medical Journal*, **i,** 685.

SCOTT, J. H. S. (1956): Assessment of Ununited Fractures of the Carpal Scaphoid. *Proceedings of the Royal Society of Medicine* (Section of Orthopaedics), **49,** 961.

SNODGRASS, L. E. (1933): End-results of Carpal-scaphoid Fractures. *Annals of Surgery*, **97,** 209.

SOTO-HALL, R. (1945): Recent Fractures of the Carpal Scaphoid. *Journal of the American Medical Association*, **129,** 335.

SOTO-HALL, R., and HALDEMAN, K. O. (1934): Treatment of Fractures of the Carpal Scaphoid. *Journal of Bone and Joint Surgery*, **16,** 822.

SQUIRE, M. (1959): Carpal Mechanics and Trauma. *Journal of Bone and Joint Surgery*, **41–B,** 210.

STEWART, M. J. (1954): Fractures of the Carpal Navicular (Scaphoid). *Journal of Bone and Joint Surgery*, **36–A,** 998.

STIMSON, L. A. (1902): Fracture of the Carpal Scaphoid with Dislocation Forward of the Central Fragment. *Annals of Surgery*, **35,** 574.

WATSON-JONES, R. (1934): Inadequate Immobilization and Non-union of Fractures. *British Medical Journal*, **i,** 937.

WATSON-JONES, Sir R. (1955): *Fractures and Joint Injuries.* Fourth edition, p. 610. Edinburgh and London: E. & S. Livingstone Ltd.

EXCISION OF THE LUNATE BONE IN KIENBÖCK'S DISEASE*

H. S. GILLESPIE, VANCOUVER, CANADA

From the Trauma Research Unit, Department of Surgery, University of British Columbia

There are different opinions on the results of excision of the lunate bone in Kienböck's disease. Dornan (1949) described sixteen cases, reporting seven excellent, four good and four fair results, with one poor result. Marek (1957) analysed four cases in detail, with three good results and one in which arthrodesis of the wrist was required later. Ståhl (1947) described fourteen cases and reported generally poor results. This paper will add a report based on twenty-four operations performed at two large Vancouver hospitals in the years 1935 to 1958 inclusive.

CLINICAL MATERIAL

Method of study—Only patients who could be examined by the author or a reliable medical witness and subsequently radiographed were included. The group consisted of eleven Workmen's Compensation Board cases, four military and nine private cases. A uniform radiological examination of the wrist was used including lateral views in full dorsiflexion and full palmar flexion, and antero-posterior views in full radial and full ulnar deviation.

General features—Twenty-two patients were men and two women. The right wrist was affected in eleven patients and the left in thirteen. The age of the patients was between twenty and forty years in twenty-one instances; in three it was between forty and fifty years. In seventeen instances the lunate bone was excised within two years of the beginning of symptoms; in three this interval was eight to fourteen years. In four patients no definite time for the onset of symptoms could be obtained. A history of accident was noted in twenty men: the most common direction of strain was dorsiflexion of the wrist, but this was quite variable. The indication for operation was always pain. All patients showed limitation of movement of the wrist.

Operation—The operation was performed by different surgeons. Twenty-one patients had dorsal approaches and three had palmar. Histologically, the lunate bone in all patients showed the typical features of Kienböck's disease with avascular necrosis of bone.

ASSESSMENT OF RESULTS

Symptoms—One patient stated that the wrist was worse after operation. The remainder stated that their wrists were improved. *Pain*—A minor ache on heavy effort was noted by most patients. Moderate to severe pain was present in three cases. It was most pronounced in ulnar deviation and dorsiflexion. *Ability to work*—One man is unable to work. Two patients returned to lighter work. The remaining twenty-one returned to their original work or to work of a similar kind.

Signs—No patient presented increased warmth over the wrist or fluid effusion at the time of examination. Soft-tissue thickening was observed in six patients. *Range of movement*—This was variable: average figures were dorsiflexion 40 degrees, palmar flexion 40 degrees, radial deviation 22 degrees and ulnar deviation 19 degrees. The diminution of ulnar deviation as compared with radial deviation was striking. *Crepitus* was noted in thirteen patients and was most marked with the wrist in dorsiflexion and ulnar deviation.

Radiographic findings. *Osteoarthritis*—Marginal lipping of the radius was observed on its dorsal aspect in six patients. This appearance did not progress or regress after operation.

* This paper was delivered at the meeting of the Canadian Orthopaedic Association, June 27–30, 1959.

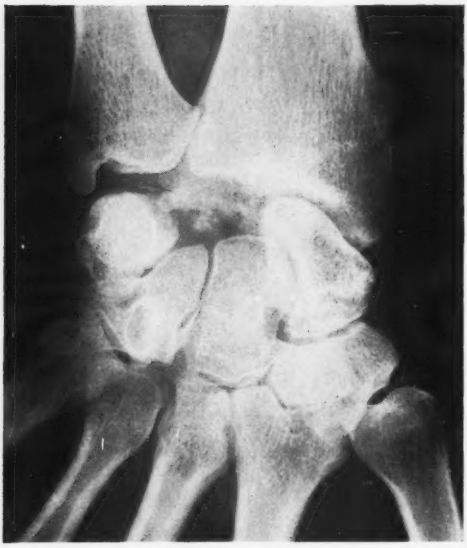


FIG. 1

FIG. 2

Case 1. Figure 1—Radiograph after operation showing normal ulnar styloid process and no evidence of cyst formation. Figure 2—Sixteen years after excision of the lunate bone. There are cystic changes in the triquetrum and the ulnar styloid process, which has altered in shape.

FIG. 3

FIG. 4

Case 2. Figure 3—Radiograph before operation showing prominent ulnar styloid process and no evidence of cyst formation. Figure 4—Ten years after operation. Excision of the carpal lunate bone was carried out eight years after diagnosis. Note the large radial exostosis. Cystic changes are present in the ulnar head and styloid process, which has altered in shape.

One patient showed narrowing of the radio-scaphoid articulation before operation; one year later this appearance had not altered. In the other patients radiocarpal or intercarpal narrowing was not remarkable.

Carpal bone shift—There was only minor carpal bone shift. The capitate moved proximally, especially with the wrist in ulnar deviation, for distances varying from one to four millimetres (Fig. 5). However, in no patient was there intimate relationship to the radius in dorsiflexion and palmar flexion. The scaphoid bone appeared to undergo a slight change in relationship to the capitate. In the normal wrist the scaphoid bone recovers from a 90 degrees volar inclination in palmar flexion to 0 degrees inclination in dorsiflexion. After excision of the lunate bone some volar inclination was maintained in dorsiflexion (Fig. 6). This led, in the antero-posterior radiographs, to an apparent foreshortening of the scaphoid bone.

Cyst formation—Apparent cyst formation either appeared or enlarged in twelve patients (Figs. 1 to 4). The triquetral bone was involved in nine patients, the lower end of the ulna in eight and the capitate bone in one case. In the lower end of the ulna the styloid process was the usual site. In four patients two bones were involved and in one patient three.

FIG. 5 FIG. 6

Case 3. Figure 5—Antero-posterior radiographs seven years after operation. There is no carpal bone shift, no arthritic change and no cyst formation. Figure 6—Lateral radiographs showing that residual volar inclination of the scaphoid is maintained in dorsiflexion.

Bone reformation—This was absent in five patients, slight in eleven, moderate in three and marked in two. There was no relationship between bone reformation and limitation of wrist motion.

Ulnar shortening—Persson (1945) described ulnar shortening as the important etiological factor in production of the Kienböck lesion. In the author's series eleven patients had two millimetres or more of ulnar shortening. This ulnar shortening appeared to show an inverse relationship to cyst formation. Ten patients with normal ulnar length and only two with ulnar shortening had cystic changes. The size and shape of the ulnar styloid process was altered in five patients. In two patients it appeared to regress in size and in three to broaden (Figs. 1 to 4).

Classification of results—The results were assessed on the basis of pain, grasp, range of movement, radiological changes and the need for a change of occupation (Table I).

In the *excellent* group there was no significant pain, grasp was of good power, range of antero-posterior movement was 70 degrees or more, range of lateral movement was 30 degrees or more, radiographic changes were absent or minimal and there had been no change of occupation (Figs. 5 and 6).

Three of the *good* results were placed in this category because of limited wrist movement. Two were included because of mild pain and minimal radiographic change.

In the *fair* group one patient had moderate pain and extensive radiographic change between the triquetrum and the ulna. The other patient had a similar amount of pain and extensive radiographic change in the radiocarpal joint.

TABLE I

RESULTS OF EXCISION OF THE LUNATE BONE (TWENTY-FOUR CASES)

Result	Number of cases
Excellent	16 (67%)
Good	5 (21%)
Fair	2 (8%)
Poor	1 (4%)

TABLE II

RELATION OF RESULTS TO LENGTH OF FOLLOW-UP (TWENTY-FOUR CASES)

Length of follow-up (*years*)	Excellent	Good	Fair	Poor
0–2	2	1	—	1
2–4	—	1	—	—
4–6	5	1	—	—
6–8	5	1	—	—
8–10	4	—	1	—
Over 10	—	1	1	—
Total	16	5	2	1

The patient placed in the *poor* category was unable to work because of severe pain seemingly originating from the radio-scaphoid articulation. Arthrodesis of the wrist is contemplated.

There was no definite relationship between the quality of the result and the period of follow-up (Table II).

Three excisions were carried out eight to fourteen years after the onset of symptoms. In one the result was good, one was fair and the third was poor.

DISCUSSION

In normal wrists the triquetrum and the lower end of the ulna come into close relationship in full ulnar deviation. This relationship was radiographically more intimate after excision of the lunate bone. Evidence suggesting that there is an abnormal contact between the triquetrum and the ulnar styloid process appeared in the following observations. Firstly, pain is most common in ulnar deviation and dorsiflexion of the wrist. Secondly, limitation of ulnar deviation is marked, indicating a possible pain-prevention mechanism. Thirdly,

cysts were commonly seen in the unusual sites of the triquetrum and the lower end of the ulna. Fourthly, cysts were more common in the patients in whom ulnar length was normal as compared with patients with ulnar shortening. Finally, changes in the size and shape of the ulnar styloid process were observed.

The pain from this contact is usually not severe. It is suggested that in unsatisfactory cases attention should be focused on this area. If the pain appears to originate at this point removal of the lower end of the ulna might be considered in order to salvage a useful wrist.

SUMMARY

1. Twenty-four cases of excision of the lunate bone in Kienböck's disease have been reviewed.
2. In general, the results have been favourable, with 88 per cent excellent or good and 12 per cent fair or poor.
3. Abnormal contact between the triquetrum and the styloid process of the ulna appeared in many cases, but this contact did not appear to prejudice the results.
4. Excision after prolonged symptoms of the disease gave disappointing results.

The work was supported by funds from the Department of National Health and Welfare, and from Johnson & Johnson Ltd., Montreal, Canada.

The author wishes to acknowledge the advice and encouragement of Dr F. P. Patterson, Director, and Dr K. S. Morton, Assistant Director, of the Trauma Research Unit, and the help of Miss Kay Hoskins in the preparation of the illustrations.

REFERENCES

DORNAN, A. (1949): The Results of Treatment in Kienböck's Disease. *Journal of Bone and Joint Surgery*, **31–B,** 518.

MAREK, F. M. (1957): Avascular Necrosis of the Carpal Lunate. *Clinical Orthopaedics*, **10,** 96.

PERSSON, M. (1945): Pathogenese und Behandlung der Kienböckschen Lunatummalazie. *Acta Chirurgica Scandinavica*, 92, Supplementum 98, pp. 51 and 79.

STÅHL, F. (1947): On Lunatomalacia (Kienböck's Disease). *Acta Chirurgica Scandinavica*, 95, Supplementum 126.

CALCAREOUS DEPOSITS ABOUT THE METACARPO-PHALANGEAL JOINTS

A. G. POLLEN, LONDON, ENGLAND

From St Mary's Hospital, London

The occurrence of calcareous deposits in the supraspinatus tendon and in the abductor of the hip is well recognised, but similar deposits about the metacarpo-phalangeal joints are relatively uncommon and have received scant attention in British publications.

Sandström (1938) reported 329 cases of calcareous deposits in various parts of the body chiefly in the shoulder and hip regions, but he could find only six cases in which the fingers were involved. He considered that these deposits were of similar origin wherever they occurred and called the condition myotendinitis calcarea (1951). Carroll, Sinton and Garcia (1955) described 100 cases from the New York Orthopaedic Hospital of deposits in the wrist, hand and fingers, the commonest site being the distal end of the tendon of flexor carpi ulnaris (thirty-seven cases): some of their cases involved the metacarpo-phalangeal joints. Cooper (1942) reported eight cases of deposits about the metacarpo-phalangeal joints, and Key (1949) reported four more. In Great Britain the only cases reported were those of Hitchcock and Langton (1959) who described six cases in the hand and fingers, three of them at the metacarpo-phalangeal joints. In this paper a further four cases will be described.

CLINICAL PICTURE

The onset of symptoms is acute, sometimes with mild pyrexia. Pain is often severe, and there may be marked swelling, heat and redness of the affected joint. There is considerable restriction of movement of the finger, due to pain. Thus the condition may simulate infective or gouty arthritis. Sometimes an acutely tender nodule may be felt near to the joint. As a rule the lesion is a solitary one.

Investigations—The erythrocyte sedimentation rate may be increased, though often it is normal. The total and differential white cell count is normal. The serum phosphorus, calcium, uric acid and alkaline phosphatase are within normal limits (Hitchcock and Langton 1959).

Radiographic features—During the acute phase a dense, discrete calcareous mass is seen at one side of the metacarpal head and adjacent to the metacarpo-phalangeal joint (Fig. 1). As the symptoms subside, the deposit becomes less dense and more diffuse (Fig. 2). Eventually the deposit disappears entirely, often within a few months.

Differential diagnosis—The importance of acute infective arthritis in the differential diagnosis of calcareous deposits must be stressed. In Hitchcock and Langton's (1959) series the initial diagnosis was incorrect in the three cases involving the metacarpo-phalangeal joints and antibiotics were given to these patients. Similarly, in the cases to be described in this paper the original diagnosis was acute infective arthritis in three of the four patients, and they were given penicillin for a few days until the significance of the radiographic appearances was appreciated.

The other important feature in the differential diagnosis is the presence of a tender nodule. This was present in two of Hitchcock and Langton's (1959) patients, and was found in one of the cases to be described in this report.

Clinical course—The acute symptoms generally persist for several days and then gradually subside. As the swelling and tenderness diminish, movement returns. A nodule may remain even after all tenderness has gone.

ETIOLOGY

There seems little doubt that these calcareous deposits are similar to those occurring in tendinous tissue elsewhere, and that the causative factors are the same. Cooper (1942) examined the lesions microscopically and found degenerative changes in fibrous tissue and some narrowing of arterial lumina. Sometimes "foreign body" giant cells were seen. He suggested that the primary lesion was degenerative, possibly from local ischaemia, and that the deposition of calcium salts occurred secondarily.

Injury does not seem to be an important factor in determining the onset. Carroll, Sinton and Garcia (1955) obtained a history of injury in only a third of their cases. Seidenstein (1950) and Cooper (1942) did not associate the lesion with trauma.

There is no evidence for an infective agent in these cases, though there is an acute aseptic inflammatory process. Antibiotics do not hasten resolution. Calcareous deposits do not appear to be related to gout, and serum uric acid levels are always within normal limits.

TREATMENT

The lesions are self-limiting and will resolve without specific treatment. Carroll, Sinton and Garcia (1955) found that in untreated patients the acute symptoms subsided in about three weeks. With simple splintage relief was obtained in an average period of nine days. Many of their patients were treated by local injection of 1 per cent procaine into the deposits: the acute symptoms subsided within a few hours. The addition of hyaluronidase, or of hydrocortisone, did not enhance the effect of the local anaesthetic. A number of patients were treated surgically by evacuation of the calcareous deposit. Key (1949) favoured local operation rather than needling, but Cooper (1942) recommended aspiration of the deposit. Other workers (Martin and Brogdon 1957, Sandström 1938, Copland and Michel 1939) have advocated radiotherapy.

Since the condition is self-limiting, simple splinting of the finger should suffice, though injection of procaine may be of value when severe pain persists.

CASE REPORTS

Case 1—A male clerk aged thirty-three years complained of severe pain and swelling at the base of the left ring finger. He had suffered this for two days, though for a week the finger had been aching a little. There was no history of any injury. Examination revealed considerable swelling over the dorsum of the left hand, related to the fourth metacarpo-phalangeal joint. There was extreme tenderness in this area, and the skin was red and tense. Movements of the finger were severely restricted, and there was some impairment of range in the adjacent fingers.

There was pyrexia of 99·2 degrees Fahrenheit. The white cell count was within normal range, with a normal differential count. The erythrocyte sedimentation rate (Westergren) was 16 millimetres in the first hour. The serum uric acid estimation was 6·5 milligrams per 100 millilitres—within the normal limits. A diagnosis of acute infective arthritis was made and treatment with antibiotics and splintage of the finger was instituted. He was given 600,000 units of procaine penicillin daily for two days.

Radiographs then showed a discrete dense rounded shadow at the radial side of the fourth metacarpal neck (Fig. 1). Penicillin was discontinued and the finger was kept splinted in a position of comfort. Within another three days the pain and swelling were much less severe, and after a further two weeks they had almost completely subsided and movements of the finger were only slightly restricted. Six weeks after the onset of symptoms the only abnormality was a small non-tender nodule in front of the fourth metacarpal head. Radiographs showed the calcification to be much less dense, and more diffuse (Fig. 2). Fifteen months

FIG. 1

FIG. 2

Case 1. Figure 1—Radiograph of left hand showing dense calcareous deposit to the radial side of the fourth metacarpal head. Figure 2—The same hand six weeks later, showing faint speckled calcification.

FIG. 3

FIG. 4

Case 2. Figure 3—Radiograph of left hand showing calcareous deposit to the radial side of the second metacarpo-phalangeal joint. Figure 4—Radiograph of the same hand five weeks later. The calcification has almost disappeared.

later the hand was clinically normal and the nodule had disappeared: radiographs showed no sign of calcification.

Case 2—A sales representative aged forty-two years complained of aching about the base of the left index finger. He gave a rather vague history of minor injury to the hand two months before. On examination there was slight swelling of the metacarpo-phalangeal joint of the index finger, with some restriction of movement. There was no pyrexia. Radiographs of the hand showed a lobulated shadow, with a proximal "tail," lying antero-laterally to the metacarpo-phalangeal joint of the index finger (Fig. 3). Two days later, while in another part of the country, the symptoms became much more severe. He was admitted to the local hospital, where he was thought to have infective arthritis and was given penicillin (250,000 units three-hourly) for three days. The finger was splinted. Thereafter he returned to London, with less pain but still with considerable swelling and restriction of joint movement. The finger was rested for a further four days, by which time the pain had almost gone. He then attended for local short-wave diathermy and movements were encouraged.

Five weeks after the onset of acute symptoms the pain had completely gone and finger movements were full. A radiograph showed only faint speckled calcification (Fig. 4). Two years later the finger was clinically normal, and radiographs showed no sign of the original calcareous deposit.

FIG. 5

FIG. 6

Case 3. Figure 5—Radiograph of right hand showing a large calcareous deposit near the fifth metacarpo-phalangeal joint. Figure 6—Radiograph of the same hand four months later showing complete absorption of the deposit.

Case 3—A woman aged seventy-four years fell and injured her right hand. Five days later she reported with severe pain about the base of the right little finger. On examination there was marked swelling over the dorsum of the fifth metacarpo-phalangeal joint, with extreme tenderness between the fourth and fifth metacarpal heads. Movements of the little and ring fingers were restricted and painful. Her temperature was 100·6 degrees Fahrenheit and the clinical picture suggested infective arthritis of the fifth metacarpo-phalangeal joint. Treatment by local splinting and procaine penicillin (600,000 units daily) was instituted.

Radiographs taken the next day showed a dense irregular calcareous deposit at the radial side of the fifth metacarpo-phalangeal joint. There were degenerative changes at the interphalangeal joints, but no bony injury (Fig. 5). Penicillin injections were continued for two days. After nine days all pain and swelling had gone and movements were fully restored. Four months later the finger was entirely normal and a radiograph showed that the calcareous deposit had been absorbed (Fig. 6).

Case 4—A male medical practitioner aged thirty-one noticed an aching pain around the third metacarpo-phalangeal joint of the left hand. Radiographs showed a bilocular dense calcareous deposit to the radial side of the third metacarpo-phalangeal joint. No treatment was prescribed because the symptoms were slight.

About six months after this mild onset, sudden severe symptoms developed. The joint became exquisitely painful, tender and swollen, and any movement of the finger exacerbated the pain. Further radiographs were taken. The calcareous deposit was still present, but its outline was not so sharply defined and it was less dense. The serum uric acid was within normal limits. The patient was at first treated for gouty arthritis with hourly doses of colchicine by mouth. No dramatic improvement in the symptoms ensued, though a severe toxic reaction followed the cessation of colchicine therapy. The pain and swelling of the joint gradually subsided in about seven days without further treatment, and complete recovery followed. A radiograph taken eleven months later showed complete disappearance of the calcareous deposit.

SUMMARY

1. Previous reports of calcareous deposits about the metacarpo-phalangeal joints are reviewed, and four further cases are described.
2. The clinical features are described, with reference to the possibility of erroneous diagnosis.
3. Conservative treatment is recommended.

I am grateful to Mr J. C. Adams for his help and encouragement in the preparation of this paper. I wish to thank Mr Rodney Sneath for giving me the details of the patient in Case 3, whom he treated. I would like to express my thanks to Dr P. N. Cardew and the staff of the Photographic Department of St Mary's Hospital for reproduction of the radiographs.

REFERENCES

AITKEN, A. P., and MAGILL, H. K. (1951): Calcareous Tendinitis of the Flexor Carpi Ulnaris. *New England Journal of Medicine*, **244**, 434.

BOSSI, R. (1954): Peritendinitis Calcarea with Multiple Foci in the Finger Joints of a Young Subject. *British Journal of Radiology*, **N.S. 27**, 692.

CARROLL, R. E., SINTON, W., and GARCIA, A. (1955): Acute Calcium Deposits in the Hand. *Journal of the American Medical Association*, **157**, 422.

COHEN, I. (1924): Calcareous Deposit at the Insertion of Flexor Carpi Ulnaris Tendon Following Trauma. *American Journal of Surgery*, **38**, 172.

COOPER, W. (1942): Calcareous Tendinitis in the Metacarpo-phalangeal Region. *Journal of Bone and Joint Surgery*, **24**, 114.

COPLAND, S. M., and MICHEL, M. (1939): Peritendinitis Calcarea. *New Orleans Medical Journal*, **92**, 261.

DE PALMA, A. F. (1947): Calcareous Deposits in Soft Tissues About the Proximal Interphalangeal Joint of the Index Finger. *Journal of Bone and Joint Surgery*, **29**, 808.

GONDOS, B. (1953): Calcification About the Wrist with Acute Pain. (Periarthritis Calcarea). *Radiology*, **60**, 244.

GONDOS, B. (1957): Observations on Periarthritis Calcarea. *American Journal of Roentgenology*, **77**, 93.

HAMILTON, A. R. (1951): Calcinosis. *Journal of Bone and Joint Surgery*, **33–B**, 572.

HITCHCOCK, E. R., and LANGTON, L. (1959): Peritendinitis Calcarea with Special Reference to the Hand. *Journal of the Faculty of Radiologists*, **10**, 86.

HITCHCOCK, H. H. (1937): Calcium Deposits About Joints. *Western Journal of Surgery, Obstetrics and Gynecology*, **45**, 353.

JANSEN, K. F. (1943): Calcareous Peritendinitis. Two Cases with Localisation to the Fingers. *Acta Radiologica*, **24**, 285.

KEY, A. J. (1949): Calcium Deposits in the Vicinity of the Shoulder and of Other Joints. *Annals of Surgery*, **129,** 737.

LAPIDUS, P. W. (1943): Infiltration Therapy of Acute Tendinitis with Calcification. *Surgery, Gynecology and Obstetrics*, **76,** 715.

MARTIN, J. F., and BROGDON, B. G. (1957): Peritendinitis Calcarea of the Hand and Wrist. *American Journal of Roentgenology*, **78,** 74.

MASERITZ, I. H. (1935): Acute Intermetacarpophalangeal Calcification. *Journal of Bone and Joint Surgery*, **17,** 1,017.

MILCH, H., and GREEN, H. H. (1938): Calcification About the Flexor Carpi Ulnaris Tendon. *Archives of Surgery*, **36,** 660.

OLDFIELD, M. C. (1951): " Tennis Thumb " Tendinitis with Calcification in Flexor Pollicis Longus. *Lancet*, **i,** 1,151.

PHALEN, G. S. (1952): Calcification Adjacent to the Pisiform Bone. *Journal of Bone and Joint Surgery*, **34–A,** 579.

SANDSTRÖM, C. (1938): Peritendinitis Calcarea. A Common Disease of Middle Life: Its Diagnosis, Pathology and Treatment. *The American Journal of Roentgenology*, **40,** 1.

SANDSTRÖM, C. (1951): Calcification of the Intervertebral Discs and the Relationship Between Various Types of Calcifications in the Soft Tissues of the Body. *Acta Radiologica*, **36,** 217.

SEIDENSTEIN, H. (1950): Acute Pain in the Wrist and Hand Associated with Calcific Deposits. *Journal of Bone and Joint Surgery*, **32–A,** 413.

SHEPHARD, E. (1955): Deposit of Calcium Salts at the Wrist. Report of Two Cases. *Journal of Bone and Joint Surgery*, **37–B,** 453.

VASKO, J. R. (1946): Calcareous Tendinitis of a Flexor Tendon of the Finger. *Journal of Bone and Joint Surgery*, **28,** 638.

WINCHESTER, J. W., and MEKIE, E. C. (1947): Tendinitis of Flexor Carpi Ulnaris. *British Journal of Radiology*, **N.S. 20,** 482.

THE HEREDITARY FACTOR IN OSTEOCHONDRITIS DISSECANS

J. STOUGAARD, SORØ, DENMARK

From the Department of Surgery, County Hospital, Sorø

Since the term osteochondritis dissecans was introduced by König in 1887 the literature has brought several examples of an inherited factor which possibly plays a part in giving rise to the disease. In certain families the incidence of osteochondritis dissecans is higher than the average, although in most patients it has been impossible to demonstrate familial occurrence. These findings might indicate that a possible inherited factor is not invariably a prerequisite or that this factor alone cannot elicit the disease.

Familial occurrence was demonstrated most convincingly by Nielsen (1933) in his series of osteochondritis dissecans affecting the elbow joint. In a study of radiographs of 1,000 normal men he found that 4·1 per cent showed evidence of osteochondritis dissecans in an elbow. Moreover, the male relatives of patients with definite radiological evidence of osteochondritis dissecans showed unmistakable signs of the disease in 14·6 per cent. His series included a family of eight brothers in which two monozygotic twins, and a third brother, had osteochondritis dissecans of the elbow. Other familial cases have been reported. Wagoner and Cohn (1931) described two families. In one family the father, son and a paternal uncle had osteochondritis dissecans of the knees; and in the other two brothers had both knees involved. Bilateral involvement was found by Bernstein (1925) in two sisters and their brother. An almost analogous case was reported by Gardiner in 1955. In the same year Pick described a family in which the mother and three of her four children had osteochondritis dissecans of either elbow or knee. Novotny (1952) found involvement of both knees in two brothers.

Multiple joint involvement in the same patient might indicate a hereditary trait—that is, reduced resistance in the joints concerned to injury. When multiple lesions exist both elbows or both knees are most often involved. Nielsen reported that in 25 per cent of his patients with osteochondritis dissecans of the elbows both were involved. Involvement of both elbows and both knees was described by Watson-Jones (1943). In the same year Wiberg reported a patient with affection of both knees, which incidentally showed spontaneous recovery. Roberts and Hughes (1950) mentioned a patient with osteochondritis dissecans of both elbows and both hips, and Hay (1950) reported one patient in whom both patellae, both capitula and radial heads were affected, and another with involvement of both elbows and one knee. Osteochondritis dissecans of both knees and one elbow was reported by King in 1935.

Thus, osteochondritis dissecans affecting several joints in one patient or in several members of the same family is not uncommon. In a follow-up study at the County Hospital, Sorø, Denmark, of forty-eight patients with osteochondritis dissecans of the elbow or knee, interrogation led us to the family reported below. In this family osteochondritis dissecans of the elbow or knee, or of both, occurred in three generations. The family comprised the grandfather, aged seventy-seven, and his eighteen direct descendants. Three of the descendants have not been radiographed, but these three had a negative history and no objective signs of osteochondritis dissecans.

The grandfather had not previously been radiographed, so the radiological diagnosis was somewhat questionable. Advanced osteoarthritis was found in both knees. In the left knee there was also a defect in the medial femoral condyle, which might be ascribed to osteochondritis dissecans, and a large loose body. The patient reported that this loose body had been present since the age of twelve. During his childhood there had been several episodes of locking, pain and swelling of the left knee; so on the basis of the history the diagnosis seems fairly

certain. His deceased wife was said to have had a tuberculous lesion of the left elbow, but further details were not obtainable. Of this marriage there were four children (all grown up).

Their eldest son gave a history very suggestive of osteochondritis dissecans of both knees. From childhood there had been complaints of pain, swelling and locking. Radiography showed the effects of osteochondritis dissecans on the medial femoral condyle, and osteoarthritis of both knees. This patient has two children, the elder of whom showed radiographic evidence of osteochondritis dissecans with a loose body (asymptomatic). The younger sister showed no sign of the disease.

The second offspring, a daughter, showed the effects of osteochondritis dissecans of both knees, with moderate osteoarthritis. From childhood there had been trouble with the right knee. The eldest of her four children had osteochondritis of the right elbow, with a loose body. The other three children—twin boys of sixteen and a girl of thirteen—showed osteochondritis of both elbows and both knees. Judging from external appearances, the twins are monozygotic. One of them had an operation on the right knee for removal of a loose body. The elbows were severely affected in this case and the capitulum had almost disappeared; its site was ragged, with several fragments in the joint space apart from actual loose body formation. In the other twin the changes were not so marked. Apart from moderate symptoms

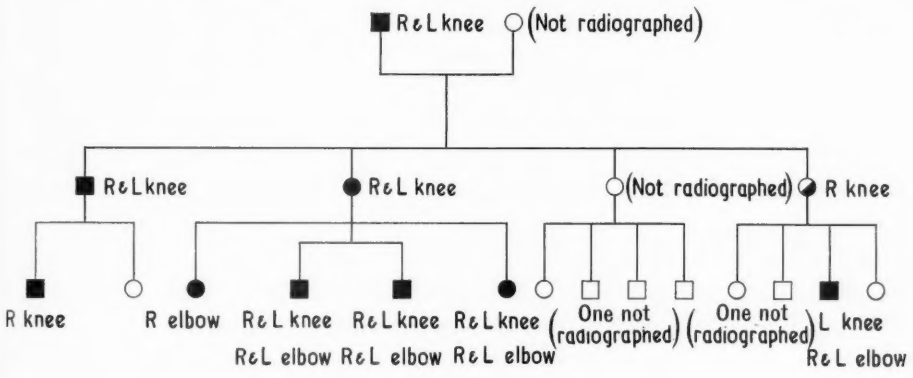


FIG. 1

Family tree. Affected members shown in solid black.

from the knees, this patient had approximately 10 degrees' limitation of extension of the left elbow and a tendency to locking. The capitula were flattened, but with smooth joint surfaces and with a loose body, the size of a pea, in each joint. If the disease is accepted as being hereditary in this family, and if the twins were in fact identical, the predisposition to osteochondritis dissecans must have been the same in each. The difference in the degree of manifestation must then be explained by external factors. The younger sister showed unmistakable signs of osteochondritis dissecans in both elbows and knees, but no prominence on the articular surface and no loose body. Her only complaint was of cracking in the left knee after prolonged use.

The next daughter was not radiographed, but the history and the objective findings gave no indication of osteochondritis dissecans. Her four children, three of whom were radiographed, had no joint trouble, and the films showed no evidence of osteochondritis dissecans.

The youngest daughter had no joint symptoms until the age of forty, when she had trouble with the right knee. Radiographs did not show osteochondritis dissecans or its effects. She had four children (three were radiographed), only one of whom is affected with involvement of both elbows and the left knee. The involvement of the knee was not discovered until the present study, because the patient had never complained about it.

The family tree summarises the examples of osteochondritis dissecans in its different members (Fig. 1). Whether the " tuberculous " lesion of the grandmother's elbow should be ascribed to osteochondritis dissecans is difficult to decide. However, it is likely that in this family there is an inherited factor governing osteochondritis dissecans and that it is transmitted as a dominant factor. This is supported by the occurrence of the disease in the presumably monozygotic twins and by the absence of any consanguineous marriages. The conclusion regarding the heredity factor in osteochondritis dissecans in this family and the apparently dominant transmission cannot be extended to apply to osteochondritis dissecans in general because the series is far too small. In addition, it must be considered selected, because it was the only family in the follow-up study to be subjected to a systematic investigation.

SUMMARY

Nine cases of osteochondritis dissecans of the elbow and knee in three generations of the same family are described. There was clear evidence of a dominant inherited factor.

The author would like to acknowledge his indebtedness to Dr Mogens Hauge of the University Institute for Human Genetics, Copenhagen, for assistance in respect to the genetic problems.

REFERENCES

BERNSTEIN, M. A. (1925): Osteochondritis Dissecans. *Journal of Bone and Joint Surgery*, **7**, 319.

GARDINER, T. B. (1955): Osteochondritis Dissecans in Three Members of One Family. *Journal of Bone and Joint Surgery*, **37–B**, 139.

HAY, B. M. (1950): Two Cases of Osteochondritis Dissecans Affecting Several Joints. *Journal of Bone and Joint Surgery*, **32–B**, 361.

KING, E. S. J. (1935): *Localized Rarefying Conditions of Bone*. London: Edward Arnold & Co.

KÖNIG, (1887): Ueber freie Körper in den Gelenken. *Deutsche Zeitschrift für Chirurgie*, **27**, 90.

LÖHR, W. (1929): Die Osteochondritis dissecans und das Gelenkmausleiden. *Zentralblatt für Chirurgie*, **56**, 2,242.

NIELSEN, N. A. (1933): Osteochondritis Dissecans Capituli Humeri. *Acta Orthopaedica Scandinavica*, **4**, 307.

NOVOTNY, H. (1952): Osteochondrosis Dissecans in Two Brothers. The Pre- and Developed State. *Acta Radiologica*, **37**, 493.

PICK, M. P. (1955): Familial Osteochondritis Dissecans. *Journal of Bone and Joint Surgery*, **37–B**, 142.

ROBERTS, N., and HUGHES, R. (1950): Osteochondritis Dissecans of the Elbow Joint. *Journal of Bone and Joint Surgery*, **32–B**, 356.

WAGONER, G., and COHN, B. N. E. (1931): Osteochondritis Dissecans. *Archives of Surgery*, **23**, 1.

WATSON-JONES, R. (1943): *Fractures and Joint Injuries*. Third edition. Edinburgh: E. & S. Livingstone Ltd.

WIBERG, G. (1943): Spontaneous Healing of Osteochondritis Dissecans in the Knee-joint. *Acta Orthopaedica Scandinavica*, **14**, 270.

MADURA FOOT

A Study of Twenty Cases

J. K. OYSTON, ROYAL AIR FORCE

From the Royal Air Force Hospital, Aden

This paper is based on a study of twenty consecutive cases of Madura foot treated in the Royal Air Force native hospital in Aden between December 1956 and August 1958. Madura foot or mycetoma pedis is a condition due to infection with fungal organisms belonging to the genera Nocardia or Madurella (Adams and Maegraith 1960). Nocardia is an aerobic actinomycete; Madurella is a true fungus. The picture of the disease described twenty years ago (Tribedi and Mukherjee 1939, Dixon 1941) is that of an emaciated patient with a foot grossly enlarged and pitted by numerous discharging sinuses (Fig. 1). The only effective treatment for such a patient is amputation, with all the disability implied by that procedure in a country where artificial limbs are not easily available and where little can be done to ensure employment for a cripple. However, only one of the patients in this series was of this very severe type; most of the others were seen when the disease was in an earlier stage and when there was an effective alternative to major amputation. The satisfactory results of treatment indicate the importance of early diagnosis.

FIG. 1

A patient with advanced Madura foot (Case 1). Note the gross enlargement of the foot and evidence of constitutional disturbance.

ANALYSIS OF MATERIAL

Age and sex—The patients were often rather vague about their exact ages; most were young adults. There were four women and sixteen men (Table I). Few regularly wore shoes or sandals.

History and clinical features—The patient usually complained of swelling of the foot. Pain was a feature in three, and discharge was the presenting symptom in three. The duration of symptoms was from one to 144 months (Table I), though the patients' vagueness about time made these figures rather speculative.

In eleven patients the left foot was affected; in eight the right. In one case the side involved was not recorded. Most commonly the forefoot was affected, and only in one case was the whole foot involved (Table I). There was involvement of regional lymph glands in one

TABLE I

ANALYSIS OF CLINICAL MATERIAL

Case number	Sex	History	Duration of symptoms (*months*)	Site of lesion	Radiological findings
1	Male	Progressive nearly painless swelling and sinuses	12	Left foot and inguinal glands	Gross bone destruction
2	Male	Swelling	2	Right foot	Periostitis in calcaneo-cuboid region
3	Female	—	—	Right foot	No abnormality
4	Female	Swelling and ulceration	3	Right forefoot	No abnormality
5	Male	Swelling and pain	7	Lateral aspect of right ankle	No bony abnormality
6	Male	Pain and discharge	8	Left great toe	Erosion of proximal phalanx
7	Male	Swelling	3	Left first intermetatarsal cleft	No abnormality
8	Female	Swelling	18	Left foot	Not done
9	Male	Swelling and aching	6	Left foot below medial malleolus	No abnormality
10	Male	Swelling	12	Left first intermetatarsal cleft	No abnormality
11	Male	Swelling	9	Dorsum of left foot	Not done
12	Male	Swelling	48	Base of second left toe	Erosion of phalanx
13	Male	Swelling	5	Sole of foot	No abnormality
14	Male	Swelling of foot and pain in groin	4	Right foot	No abnormality
15	Male	Swelling	10	Sole of right foot	Not done
16	Male	Swelling and some pain	144	Left second toe	Not done
17	Male	Swelling	12	Sole of left foot	Not done
18	Female	Swelling	8	Right forefoot	No abnormality
19	Male	Swelling	—	Right foot	Not done
20	Male	Lump on foot	1	Lateral aspect of left tarsus	Not done

patient. In this case necrosis and discharge occurred and the fungus was recovered from the discharging gland. In one patient the regional lymph glands were simply enlarged in consequence of secondary infection of the primary lesion.

Appearance of primary lesion—Six patients showed ulceration or the typical sinuses with raised margins (Fig. 2). In one other case the swelling was topped by several hard nodules which seemed about to break down. The other patients showed swellings varying in shape and size and in degree of attachment to the surrounding tissue. Some were fluctuant and some seemed to be multilocular.

Radiological appearances—In eight cases the radiographs showed no abnormality of bone. In three the appearances were of superficial affection of bone and in one (Fig. 3) there was obvious evidence of extensive involvement.

Systemic reaction—The lesion is essentially a local one and does not usually cause any constitutional disturbance. Only three patients were pyrexial and in all of these the pyrexia seemed to be due to secondary infection rather than to the disease itself. In none of the seven patients who had the investigation was there a leucocytosis.

FIG. 2
Gross involvement of the whole foot showing the fusiform swelling and the sinuses with raised margins (Case 1).

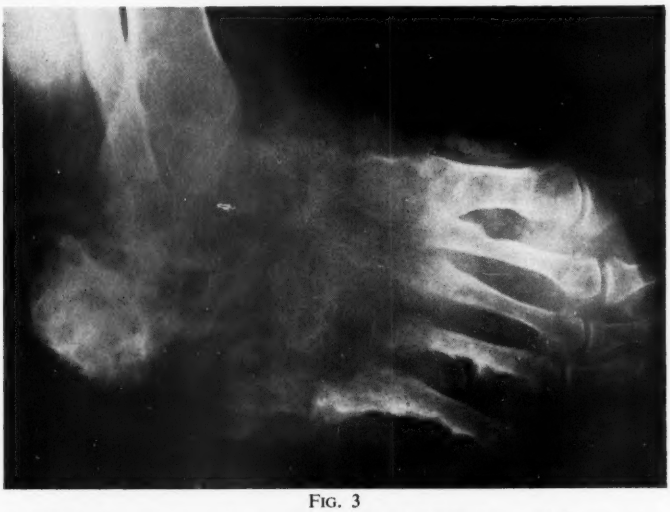


FIG. 3
Radiological appearances in a case of advanced infection. Note the gross destruction of bone and the swelling of soft tissues (Case 1).

Pathological features—The tissues involved were chiefly skin and subcutaneous tissue (eight patients) but in four muscle and tendon were affected, in three periosteum and in one bone. In four cases the tissue involved was not recorded. The recognition of the different types of causal organism (Erikson 1935, 1953, Mackinnon and Artagaveytia-Allende 1956) is a

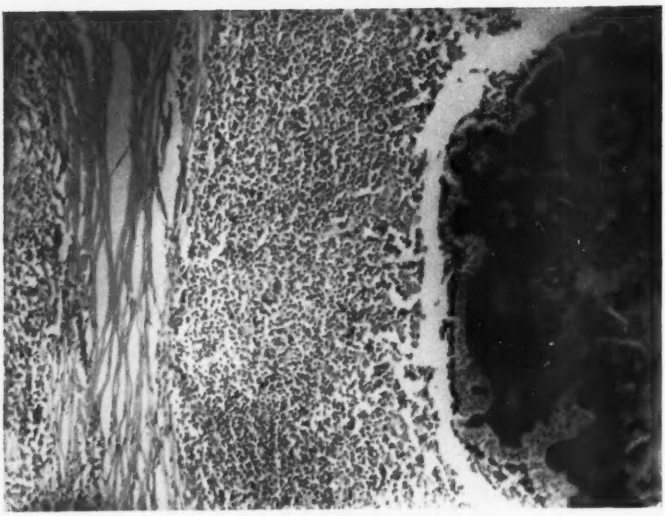


FIG. 4

Section through a lesion, showing the mycelial mass (*on the right*) surrounded by polymorphs, plasma cells and giant cells. The whole is surrounded by a fibrous " capsule " (*on the left*). (×40.)

FIG. 5

Advanced mycetoma involving forefoot. Suitable for Syme's amputation.

matter for an expert mycologist. The simplest classification—that depending on the colour of the granules present in the discharge—was accordingly used. The granules were black in three cases and yellow in nine. Their colour was not recorded in the other cases.

In the early stages there is an infected area of varying size and shape bounded by a fibrous capsule of varying thickness. This capsule may break down and further spread may thus occur. The lesion consists of aggregates of granules surrounded by inflammatory tissue. With breakdown of the capsule and involvement of other tissues the surrounding fibrous reaction becomes more intense.

Microscopically the lesion consists of a spherical collection of mycelium surrounded by cells of varying types. In some cases they are polymorphs, in others epithelioid cells. Plasma cells and giant cells may also be present. Surrounding the cells is an area of fibrous tissue (Fig. 4). In some cases there is a well marked palisade layer of eosinophil cells, adjacent to the mycelium.

TREATMENT

All the patients were treated with chlortetracycline 250 milligrams six hourly, combined with diaminodiphenylsulphone 50 milligrams daily for the first week, 100 milligrams daily for the second week and so on up to 250 milligrams daily. These drugs were not given over long periods because of doubts about their efficacy and because of the unwillingness of patients to stay long in hospital without marked evidence of improvement. They were used as adjuvants to operative treatment.

Operative treatment aimed at the removal of all infected tissues and closure of the ensuing wound. In some cases the disease area seemed to be encapsulated and could be shelled out; in others it had to be excised with a margin of healthy tissue. Amputation was only necessary when the affection was very extensive. After local excision primary closure was in one case possible; in eleven the area of skin excised was so large that the wound had to be left open to be covered later by a free graft. One patient was treated by excision of the lesion without primary closure or secondary grafting. Four patients with advanced disease had amputations; in two of them, with involvement of the forefoot only, Syme's method was used (Fig. 5). Three patients were treated by drugs alone, but unfortunately did not attend for review, and one left hospital after only a few days' treatment (Table II).

RESULTS

Unfortunately no adequate long term observation of these patients was possible because the patients simply failed to attend the follow-up clinic.

Medical treatment failed completely for similar reasons—in all three cases where it was attempted the patient either refused to stay in hospital or failed to attend the out-patient clinic.

Treatment by excision gave good results over the period during which observation was possible (Table II). In all eleven patients treated by local excision and later split-skin grafting the wounds were soundly healed at the time of discharge from hospital. The same satisfactory results followed the other methods of surgical treatment. One patient (Case 1) was kept under observation for twelve months; in this case there was advanced disease, but there was no evidence of recurrence during the period of observation.

CASE REPORT

This illustrates advanced disease of the classical type, but the case is not typical of those reported here.

A man aged about thirty years (Case 1) came to the clinic in September 1957 walking unshod with the aid of a stick. He complained of progressive swelling of the left foot over the past year and of a swelling in the left groin present for five months. There had been no pain.

TABLE II
TREATMENT AND RESULTS

Case number	Treatment	Result	Time in hospital (*days*)
1	Amputation and excision of glands	Primary healing. No recurrence ten months after discharge	83
2	Excision skin, muscle and periosteum. Free graft seventeen days after excision	90 per cent take of graft. No recurrence at one month after discharge	42
3	Excision skin and subcutaneous tissues. Free graft seven days after excision	90 per cent take of graft	61
4	Syme's amputation	Satisfactory stump	51
5	Medical only	No improvement	28
6	Excision skin, subcutaneous tissue and periosteum. Free graft sixteen days after excision	Complete take of graft	51
7	Excision skin and soft tissues	Wound healed thirty days after excision. No recurrence two months after discharge	37
8	Excision of skin and soft tissue. Free graft twenty-one days after excision	Complete take of graft	61
9	Excision of skin and soft tissue. Free graft eighteen days after excision	50 per cent take of graft. Healed thirty days after grafting	57
10	Excision of skin and soft tissue. Free graft twelve days after excision	75 per cent take of graft. Healed nineteen days after grafting	40
11	Excision of skin and soft tissues. Primary closure	Satisfactory healing	20
12	Disarticulation of toe. Free graft fourteen days after amputation	Healed in fourteen days	36
13	Excision of skin and soft tissues. Free graft twenty-eight days after excision	30 per cent take of graft. Healed fourteen days after grafting	54
14	Excision. Free graft fourteen days after excision	95 per cent take of graft	31
15	Medical only	Left hospital refusing further treatment	6
16	Medical only	Failed to attend for follow-up	27
17	Excision of skin and subcutaneous tissues. Free graft seventeen days after excision	50 per cent take of graft. Wound healed twelve days after grafting	33
18	Syme's amputation	Delayed healing. Final result satisfactory	70
19	Excision of skin, subcutaneous tissue and muscle. Free graft fourteen days after excision	Complete take of graft (length of stay because of associated epididymitis)	82
20	Excision of skin and subcutaneous tissue. Free graft fourteen days after excision	Good result	38

The patient was clearly in poor general health. Temperature and pulse were normal. The edge of his spleen was palpable two and a half inches below the left costal margin. The left foot was swollen to twice the size of the healthy one and was covered with sinuses with raised margins. The popliteal lymph glands were enlarged. There was an abscess in the left inguinal region with numerous sinuses and enlarged matted lymph glands (Fig. 6). Haemoglobin was 46 per cent; white blood cells were 5,000 per cubic millimetre—neutrophils 47 per cent, lymphocytes 51 per cent and eosinophils 2 per cent.

Radiological appearances—There was advanced destruction of bone in the middle of the foot. In spite of this extensive destruction the bony arch was well preserved (Fig. 7). There was general decalcification of the forefoot with periostitis affecting particularly the fourth and fifth metatarsal bones. The calcaneum showed numerous spherical areas of decalcification surrounded by sclerosis.

Treatment—Chlortetracycline (250 milligrams six-hourly), diaminodiphenylsulphone (50 milligrams daily for one week, 100 milligrams daily for one week and 150 milligrams daily for one week) and potassium iodide (30 grains daily) were given, together with "Imferon" (250 milligrams on alternate days) and Folic acid (5 milligrams daily).

After three weeks' treatment the patient complained of pain in the foot and groin. The swellings had softened and white blood cells were 9,100 per cubic millimetre (neutrophils 86 per cent). There was still a marked anaemia. It was decided that operation was necessary.

FIG. 6
Inguinal region showing enlarged lymph glands and sinuses (Case 1).

Below-knee amputation with primary closure over drainage tubes was done. Removal of the inguinal lymph glands was attempted but had to be abandoned before it was complete because of profuse bleeding and the patient's poor general condition. After transfusion of a pint of blood—all that could be obtained—and further treatment with iron the patient's condition gradually improved and his haemoglobin rose to 79 per cent. In November 1957 a second operation was done, the popliteal glands and the remaining inguinal glands being removed. All wounds healed primarily and the patient left hospital twelve weeks after admission. He was seen two months after discharge when there was no sign of recurrence, and ten months later was reported to be in good health.

Pathological examination—There was extensive involvement of the midtarsal region where the bone had been replaced by confluent spheres of dense fibrous tissue of an average diameter of 8 millimetres filled with caseous material off-white in colour. Islands of apparently unaffected bone were present in the forefoot and toes and the talus was only involved on its inferior surface. The calcaneum was pitted with small foci of caseation lacking a dense fibrous capsule (Fig. 8). The tibia seemed to be healthy.

On microscopy each lesion was seen to consist of a mycelial mass surrounded by polymorphs, epithelioid cells, plasma cells and giant cells. This was surrounded to a varying extent by fibrous tissue, this reaction being least marked in the early bony lesions.

The inguinal mass was formed by inflammatory cells surrounding mycelia.

DISCUSSION

It is probable that the organisms, present in the soil, gain entry through cuts and abrasions in the unshod foot. The disease affects the active man in the place most often subjected to minor cuts and in contact with the soil. The forefoot is most often affected, possibly because

FIG. 7
Radiological appearances showing preservation of bony arch in spite of gross destruction of bone (Case 1).

FIG. 8
Section through the amputated foot showing the extensive involvement and replacement of normal tissues by the mycetoma (Case 1).

it has areas of relatively soft skin between the hard weight-bearing areas. The incidence was related to occupation by Bocarro (1893) but more recently Peters (1945) showed that it depended more on whether the patient went barefoot.

Most of the patients in this series had the disease recognised while it was in an early stage. The patient whose case is described had very advanced disease and—an unusual feature—infection of regional lymph glands by the fungus. This patient showed well-marked radiological changes.

The fungus seemed to grow more readily in subcutaneous and areolar tissues and only to invade deep structures late in the course of the disease. This finding is at variance with that of Dixon (1941) who described the lesion as burrowing indiscriminately through all tissues.

Surgical treatment by local excision was generally successful; there is no reliable evidence from this study of the value of chlortetracycline and diaminodiphenylsulphone. Treatment with sulphonamides (Dixon 1941, Peters 1945) was not tried.

SUMMARY

1. Twenty cases of mycetoma pedis are described.
2. Most patients had the disease in an early stage and were cured by simple excision.
3. The importance of early diagnosis and treatment in order to avoid amputation is emphasised.

I wish to thank the Director General Medical Services Royal Air Force for permission to publish this paper, and also Air Commodore L. M. Crooks, *O.B.E.*, M.S., F.R.C.S., R.A.F. Consultant in Orthopaedic Surgery, for his advice and help.

REFERENCES

ADAMS, A. R. D., and MAEGRAITH, B. G. (1960): *Clinical Tropical Diseases*. Second edition, p. 228. Oxford: Blackwell Scientific Publications.

BOCARRO, J. E. (1893): An Analysis of One Hundred Cases of Mycetoma. *Lancet*, **ii,** 797.

DIXON, J. M. (1941): Sulfanilamide Therapy in Madura Foot. *Virginia Medical Monthly*, **68,** 281.

ERIKSON, D. (1935): The Pathogenic Aerobic Organisms of the Actinomyces Group. *Medical Research Council Special Report Series No.* 203. London: His Majesty's Stationery Office.

ERIKSON, D. (1953): Variation of Mycelial Pattern in Sporogenous and Asporogenous Actinomycetes. In *Symposium on Actinomycetales, Sixth International Congress of Microbiology*, p. 102. Rome: Fondazione Emanuele Paterno.

MACKINNON, J. E., and ARTAGAVEYTIA-ALLENDE, R. C. (1956): The Main Species of Pathogenic Aerobic Actinomycetes Causing Mycetomas. *Transactions of the Royal Society of Tropical Medicine and Hygiene*, **50,** 31.

PETERS, J. T. (1945): A Clinical Cure of Madura Foot. *American Journal of Tropical Medicine*, **25,** 363.

TRIBEDI, B. P., and MUKHERJEE, B. N. (1939): Actinomycotic and Mycotic Lesions, with Special Reference to "Madura Foot." *British Journal of Surgery*, **27,** 256.

ACETABULAR DYSPLASIA

The Acetabular Angle

IAN K. SHARP, HULL, ENGLAND

Congenital dysplasia of the hip joint, one of the common causes of osteoarthritis of the hip, has as its features deformity of the femoral head, anteversion of the femoral neck, coxa valga and varying degrees of shallow acetabulum (Lloyd-Roberts 1955). Of these, the last is the commonest and probably the most important. The etiology of hip dysplasia has been discussed by Wiberg (1939) and Hart (1952). Various estimates of the frequency of the condition have been made. Lloyd-Roberts (1955) found that 20·9 per cent of arthritic hips were initially dysplastic, Wiberg (1939) reported 25·7 per cent while Gade (1947) attributed no less than 47·9 per cent of his series to this congenital deformity. This difference in estimates is probably due to the difficulty in identifying the primary etiological factor in advanced osteoarthritis.

Wiberg (1939) evolved a radiographic measurement, the C/E angle, in his studies of congenital subluxation of the hip. This C/E angle is the angle at the intersection of two lines radiating from the centre of the femoral head (C), one passing through the point (E) at the outer edge of the acetabular roof, the other vertical to the transverse axis between C and the point C_1 at the centre of the contralateral femoral head. This angle is a means of locating the femoral head relative to the acetabulum, and also, where the femoral head is not displaced, provides an indication of the degree of development of the acetabular roof. During the course of a study of acetabular dysplasia in the adult, it became clear that the C/E angle did not provide an accurate measurement of this condition for the following reasons: 1) the centre point of a deformed femoral head cannot be located accurately; 2) subluxation, or simple loss of joint space, alters the C/E angle, giving false readings; and 3) subluxation of the contralateral hip also affects the C/E angle.

It is strictly correct to consider the entire hip joint in a study of dysplasia. Nevertheless, the feature which is constantly present is the underdevelopment of the acetabular roof, where the surface available for weight bearing is smaller than normal and therefore subject to proportionately greater pressure. A simple measurement of the acetabulum alone, without reference to the head of the femur, appears to provide a logical and possibly more accurate approach.

THE ACETABULAR ANGLE

In the radiograph of the normal acetabulum only two points lend themselves readily to mensuration, namely the lateral edge of the acetabular roof and the inferior tip of the U-figure or " pelvic tear drop." Using these two points and the horizontal line between the U-figures, the angle of inclination of the acetabulum can be measured—the acetabular angle. Of the components of this measurement, the lateral edge of the acetabular roof is the more obvious. It must be remembered, however, that this is not a fixed point on the skeleton but is in fact the radiographic projection of a ridge of bone running antero-posteriorly, more or less parallel with the x-ray beam. Similarly, the U-figure is the end-on projection of a bony ridge, the floor of the acetabular fossa. Werndorff (quoted by Wiberg 1939) made the U-figure disappear by sawing away this bony plate. Vare (1952) succeeded in outlining it by means of a flat strip of lead foil applied to the acetabular fossa and curving round the inferior margin of this, up over the inner wall of the lesser pelvis as far as the ilio-pectineal eminence. It follows that the position of the two points from which the acetabular angle is measured must vary on the radiograph with any pelvic tilt. To discover the degree of error, strips of lead foil were applied to the acetabula of a macerated pelvis to outline the U-figure as described by Vare and films were taken with

the pelvis in various positions. The tube-film distance was 100 centimetres and the beam was centred 5 centimetres cephalad to the symphysis pubis, which is a standard technique for pelvic radiography.

The views taken were: 1) Symphysis and both anterior superior iliac spines in the same horizontal plane. 2) Anterior superior iliac spines level, symphysis high: pelvis tilted forward 15 degrees. 3) Anterior superior iliac spines level, symphysis low: pelvis tilted backward 15 degrees. 4) Symphysis level with mid-point of interspinous line: right anterior superior iliac spine farther forward than left: pelvis tilted 10 degrees to left. 5) As in position 4, but with pelvis tilted 10 degrees to right.

TABLE I

THE VARIATION DUE TO RADIOGRAPHIC DISTORTION

Position	Acetabular angle (*degrees*)	
	Right	Left
1	36	40
2	37	41
3	35	39
4	38	37
5	34·5	40

The results are shown in Table I. It will be seen that the variation in the angles resulting from what would be, in clinical radiography, extremely bad positioning is no more than 4 degrees. The second and third positions are, of course, also reproducible by aiming the beam above or below the correct point 5 centimetres above the symphysis. Since the tube-symphysis distance is approximately 70 centimetres, in order to produce the effect of 15 degrees pelvic tilt it would be necessary to centre the beam nearly 20 centimetres above or below the correct point. For measurement, any film so badly centred, or in which there was obvious gross lateral tilting, would be discarded.

In order to check this further, three patients had pelvic views taken in positions 4 and 5, and in each case the acetabular angle varied less than 2 degrees. Furthermore, among the normal series were a few patients who had several films taken of the pelvis, often at considerable intervals, and in these the acetabular angles were absolutely constant, showing how well standardised is the technique, even in the hands of different radiographers.

THE NORMAL RANGE

Wiberg (1939) considered that only hips which were functionally blameless could justifiably be included in a normal series. He therefore measured his angle on the radiographs of fifty men and fifty women whose hip joints were clinically normal. These subjects were between the ages of twenty and thirty-five, and he admitted that they might include mal-developed acetabula which had not yet produced symptoms. This objection cannot be dismissed; a radiological measurement would be better to have a radiologically determined base-line.

A good hip joint should last a man throughout his lifetime. Nevertheless, for purely practical reasons an arbitrary lower age limit of sixty was chosen and from the files of the Radiology Department films of fifty male and fifty female pelves with no radiological evidence of osteoarthritis were measured, making a total of 200 hip joints. Apart from the rejection of obviously distorted views no selection was made, the films having been taken for various reasons—fracture or suspected fracture, suspected pelvic metastases, etc.

The normal range of acetabular angle is shown in Figure 1. The angle was found to be essentially the same in men and women, a surprising finding in view of the well known sex difference in the pelvic conformation. It will be seen from the graph that the normal angles fall within a narrow range. The variation in normals is demonstrated in Figures 2 to 4, where the angles are respectively the lowest, the average and the highest in the series: Figure 5 illustrates the angle in a typical congenital subluxation. Figures 2 and 3 need no further

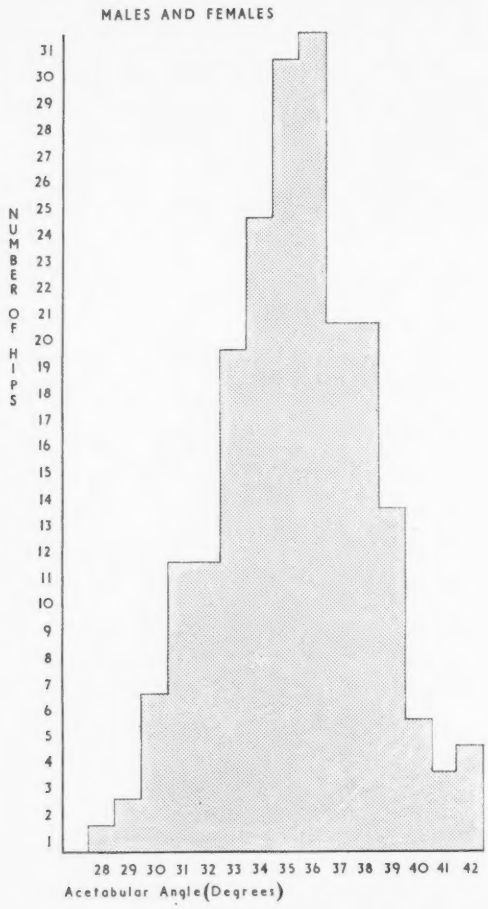


FIG. 1
The angles in the 200 hip joints aggregated.

consideration, but Figure 4 illustrates a pair of acetabula with angles of 42 degrees. It will be seen that the depth of these is such that the relatively small femoral heads are adequately supported. This demonstrates the major theoretical objection to the validity of the acetabular angle, that it does not measure the depth of the acetabulum. In practice, however, the angle of slope of the acetabulum appears to bear a fairly constant relationship to its depth, and this simple measurement provides an excellent index of the degree of acetabular development.

FIG. 2

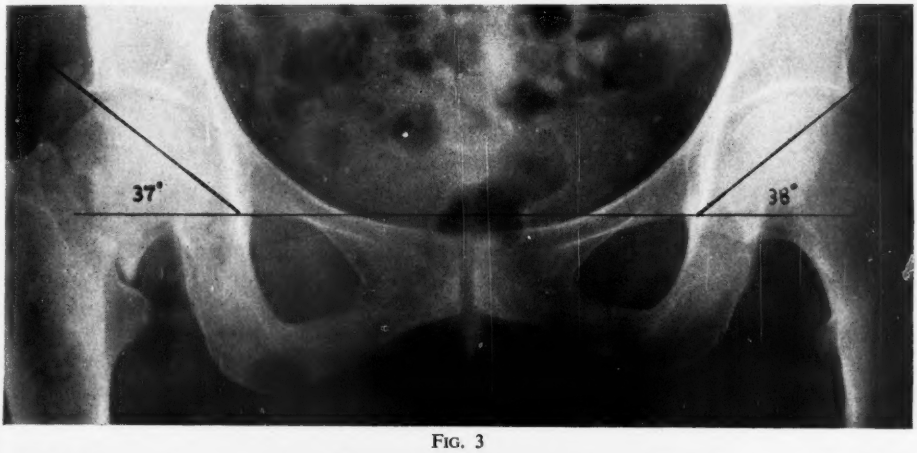


FIG. 3

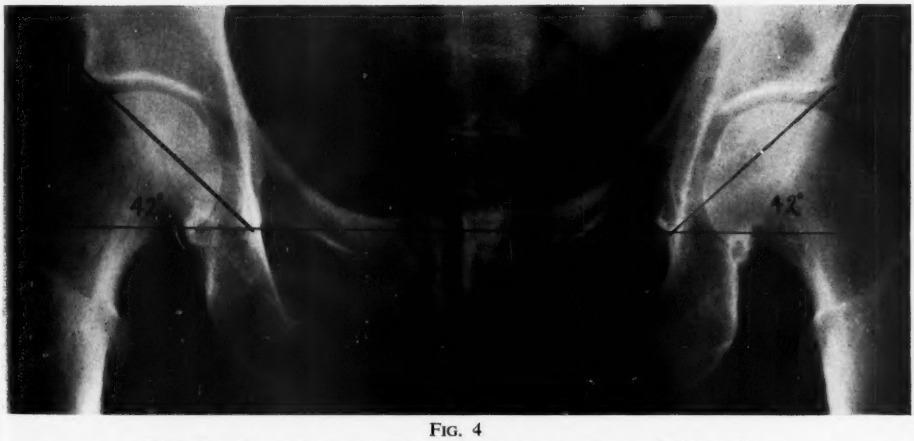


FIG. 4

Three films from the normal series illustrating low, average and high angles.

FIG. 5
Congenital subluxation of the hip. Shenton's line is just broken and the acetabular angle is 47 degrees.

SUMMARY AND CONCLUSIONS

A simple method of measuring the degree of acetabular development in the radiograph of the adult pelvis is described and arguments for its validity are advanced. This measurement is referred to as the acetabular angle. The normal values for this angle are between 33 and 38 degrees. Angles below 32 degrees are uncommon and probably of no clinical significance, whereas angles from 39 to 42 degrees are in the upper limit of normality. An angle of 47 degrees is shown in a hip with congenital subluxation. The prognosis for hip joints with acetabular angles between 42 and 47 degrees is under investigation.

REFERENCES

GADE, H. G. (1947): A Contribution to the Surgical Treatment of Osteoarthritis of the Hip-Joint. *Acta Chirurgica Scandinavica*, **95,** Supplementum **120.**

HART, V. L. (1952): *Congenital Dysplasia of the Hip Joint and Sequelae.* Springfield, Illinois: Charles C. Thomas.

LLOYD-ROBERTS, G. C. (1955): Osteoarthritis of the Hip. *Journal of Bone and Joint Surgery*, **37–B,** 8.

VARE, V. B., Jun. (1952): The Anatomy of the Pelvic Tear Figure. *Journal of Bone and Joint Surgery*, **34–A,** 167.

WIBERG, G. (1939): Studies on Dysplastic Acetabula and Congenital Subluxation of the Hip Joint. *Acta Chirurgica Scandinavica*, **83,** Supplementum **58.**

ENGELMANN'S DISEASE

Report of a Case with a Review of the Literature

EVAN A. LENNON,* BRISBANE, AUSTRALIA

MANNIE M. SCHECHTER and RICHARD W. HORNABROOK,
NEW YORK, UNITED STATES OF AMERICA

From the National Hospital for Nervous Diseases, Queen Square, London, England

Engelmann in 1929 reported a " new " disease, which he called osteopathia hyperostotica sclerotisans multiplex infantilis, and which he believed to be connected with a disturbance of the haemopoietic system. A boy, eight years of age, had complained of pains in the legs for four years. He had a short neck and trunk, long limbs, weak musculature, lumbar lordosis, flat feet and a waddling gait. There was palpable swelling of the long bones of the upper limbs. The spleen was enlarged and the testicles were small. Examination of the blood showed lymphocytosis, slight monocytosis and eosinophilia. Radiographs showed symmetrical elongation and slight curving of the long bones with thickening and sclerosis of the diaphyses. The epiphyses and joints were normal. There was sclerosis of the frontal bone and skull base.

Sear (1948) described a similar case of bone disease, and drew attention to Engelmann's paper. He gave the condition the eponymous title of Engelmann's disease.

Before Engelmann's description Cockayne (1920), at a meeting of the Royal Society of Medicine, had presented a case for diagnosis, which Fairbank (1951) subsequently recognised as probably belonging to this syndrome. Despite a thorough search through the National Registry in London, no record of this patient could be found. If still alive the patient would be forty-eight years old.

Camurati (1922) reported " a rare case of hereditary symmetrical osteitis of the lower limbs " in a boy aged seven. The clinical features and radiographic appearances resembled those of Engelmann's patient. The fifty-five-year-old father of this boy also had a history of leg pains in youth, and radiographs showed an increase in density and thickening of the shafts of the bones in both lower limbs. Seven other members of the father's family in four generations had a similar history, but were not investigated.

A number of similar patients have been reported; so the natural history of the disorder is now identifiable. Excluding the patient we are reporting, fifty-one others have been traced.

Other authors followed Camurati in drawing attention to the familial nature of this disorder. Lauterburg (1931) described " generalised familial sclerosis " in a boy aged sixteen years and in his father aged forty-two. Cocchi (1951) identified these two patients as belonging to Engelmann's syndrome and he pointed out that they had been under observation for twenty years. Ortolani and Castagnari (1953) described the condition in two siblings. Ribbing (1949) reported a family of six siblings, four of whom in the third decade showed increased density and widening of the diaphyses of the femora and tibiae; he said that Feddema found five siblings affected in the fifth and sixth decades of life. Paul (1953) described a man of twenty with this condition. His brother aged thirty-five and infant son had the same abnormalities. Jammes, Seruz, Prouxet and Duclos (1953) reported a case in a man aged twenty-two years. A brother and a sister were also affected. Buchem, Hadders and Ubbens (1955) also described a familial case. Roth (1957) reported an eighteen-year-old youth whose father and fourteen-year-old brother were similarly affected.

Sporadic cases without family history have also been reported by Fritsch (1933), Riley and Shwachman (1943), Neuhauser, Shwachman, Wittenborg and Cohen (1948), Wiedemann

* Holder of Thomas Baker Memorial Fellowship.

(1948), Michaelis (1949), Casuccio (1949), Bingold (1950), Stronge and McDowell (1950), Gvozdanović (1950), Fairbank (1951), Le Bien and Heilman (1951), Gulledge and White (1951), Gillespie and Mussey (1951), Lavine and Koven (1952), Anderson (1953), Jammes *et al.* (1953), Perassi (1954), Weingraber (1954), and Singleton, Thomas, Worthington and Hild (1956).

Griffiths (1956) described a single patient and reviewed twenty-two previously reported cases. He also drew attention to the radiographs of a patient with this disorder published by Geschickter and Copeland (1949) which is referred to in the text as of an unclassified juvenile bone dystrophy. Most of these were diagnosed in childhood, although sometimes symptoms first appeared in adults. Thus Michaelis (1949) reported the disease in a woman twenty-nine years old, and Stronge and McDowell's (1950) patient did not develop symptoms until he was twenty-eight years old.

Griffiths (1956), Anderson (1953), Jammes *et al.* (1953) and Perassi (1954) all reported the condition in adults, and Buchem *et al.* (1955) in two siblings, both of whom developed symptoms in middle life.

The range of bones involved has gradually been extended, and the evolution of the disease has become clearer. Fritsch (1933) reported a woman, thirty years old, whom he had had under observation for ten years. When first examined in 1923 she gave a history of having been frail as a child, and of having pain in the right leg for six years. There was a soft, tender swelling over the right tibia, and the radiographs showed a cortical thickening with diminution in the marrow space in the diaphysis. Later her left leg and arms became painful, with progressive changes showing in the radiographs; by 1932 the femora, tibiae, humeri and the anterior and middle cranial fossae were also involved.

Ribbing's (1949) family of six siblings developed increased density and widening of the diaphyses of the femora and tibiae. Three of these patients had leg pains, but another had no symptoms. Ribbing pointed out that the disease was progressive, but appeared to be self-limiting.

Gvozdanović (1950) described a three-month-old girl, who died of anaemia, and in whom radiological changes were very extensive, and even involved the ribs, the clavicles and the vertebrae.

Le Bien and Heilman (1951) had one patient with radiological changes in the metatarsals. Anderson (1953) is the only author who has seen changes extending into the head of the femur. Paul (1953) described a man who was first seen in 1941, at the age of twenty, complaining of swelling over the right tibia, which showed a fusiform swelling in the radiograph. In 1948 the patient complained of swelling in the left leg, and it was found that both the tibiae, the femora, and the right radius had the radiological appearances of Engelmann's disease. His family were then investigated, and his brother, aged thirty-five, whose only complaint was of episodes of double vision, was found to have typical radiological changes in both tibiae, the left femur and the left ulna. He also had a left-sided otosclerosis. Jammes *et al.* (1953) reported a man aged twenty-two, who had the history, physical signs and radiological changes of Engelmann's disease. This man had asymmetry in the length of the limbs, with the right arm longer than the left, and the left leg longer than the right. Jackson, Hanelin and Albright (1954) reviewed two patients previously described by Neuhauser *et al.* (1948) in the light of further observations. They drew attention to the fact that sometimes two separate diaphysial foci of bone expansion were seen in the femora with the intervening shaft less intensely involved or almost spared. Caffey (1956) saw that an area of bone in the region of the nutrient foramen was spared. Weingraber (1954) found that the newly formed periosteal bone in the long bones of a boy of four showed an " onion peel " structure which quickly became consolidated with the shaft, which increased three millimetres in thickness in two months.

Riley and Shwachman (1943) published two cases of " unusual osseous disease with neurological changes " in a boy and girl both aged five. The neurological features were hyperactive deep tendon reflexes and an inconstant ankle clonus. Other neurological findings,

of diplopia and otosclerosis, were reported by Paul (1953). The description of a waddling gait is common. Buchem *et al.* (1955) reported a man of forty-eight, who gave an eleven-year history of an enlarging head, failing vision, increasing deafness, occasional vertigo, stabbing headaches and muscle weakness. He was well built, with a high forehead, a prominent chin, slight enophthalmos, bilateral optic atrophy, poor thoracic movement and a dry flaky skin. Le Bien and Heilman (1951) found that the skin over the extremities felt hard and woody, as in scleroderma, and the man described by Buchem *et al.* (1955) had a hard, dry flaky skin. Some suggestion of endocrine insufficiency has been made. Thus Jammes *et al.* (1953) reported a slight decrease of 17-ketosteroid excretion in their patient. The woman reported by Buchem *et al.* (1955) had scanty axillary and pubic hair. Later she died of broncho-pneumonia but, apart from multiple subperiosteal excrescences on the bones and the widening of the cortices and diaphyses, there were no significant abnormalities.

CASE REPORT

A thirty-four-year-old man was admitted to the National Hospital for Nervous Diseases, under the care of Dr S. P. Meadows, complaining of headaches for five years and deafness for three months. An older brother of the patient had been treated for epilepsy and mental deterioration in the same hospital when an infant, and bossing of the frontal bones was remarked upon at the time. This brother died at home, aged twenty-one years, in status epilepticus, and necropsy was not performed. There were no other siblings and the parents and their relatives had had no nervous, mental or skeletal disease.

The patient was believed to have developed normally until the age of eighteen months, but when he started to walk his gait was abnormal, being shuffling and awkward. At ten years of age enlargement of the head was obvious and his limbs were of a " peculiar " shape. He was considered too delicate to go to school and consequently was largely self-educated. All his life he had been liable to attacks of epistaxis.

For five years before his admission he had been troubled by episodes of vertigo culminating on several occasions in brief disturbances of consciousness; over the same period occipital headaches, gradually increasing in frequency, made their appearance. Eventually an almost continuous generalised throbbing headache forced him to seek medical advice.

He had a massive enlargement of the head, the greatest circumference being twenty-seven and a half inches. The forehead was high and prominent. The small face was juvenile in appearance with a marked proptosis. The long, thick limbs were in striking contrast to the small hands and feet. The right arm was longer than the left. The arms had a peculiar cylindrical appearance due to the lack of musculature combined with an increase in width of the bones (Figs. 1 to 3).

There was thoracic kyphosis, marked lumbar lordosis and valgoid flat foot. The arm span was seventy-two and a half inches and the height seventy inches. The circumference of both the upper arm and forearm was nine and a half inches. The skin was thick, rough and dry. There was no beard or axillary hair and very little pubic hair. He was intelligent but the voice was quiet and juvenile. The external genitalia were infantile. The gait was stiff and shuffling, suggesting a toy soldier, and he was unable to pronate and supinate the forearms. All the deep reflexes were brisk and there was generalised muscular weakness. There was no disorder of sensation; the plantar responses were flexor. The blood pressure was 180/100 millimetres of mercury and remained consistently elevated. He had severe dental caries (Fig. 13).

Neuro-otological findings (Dr C. S. Hallpike)—There was a suggestion of eighth nerve involvement by bony changes; there was bilateral deafness, that of the left ear being perceptive and of the right conductive. There was spontaneous vestibular nystagmus to the left and the left caloric responses were abolished and the right reduced.

Ophthalmological findings (Mr Keith Lyle)—There was optic atrophy on the right. The left disc was pale but probably normal.

FIG. 1

FIG. 2

FIGS. 1 AND 2

Figure 1—The clinical appearance, showing the enlarged head, prominent forehead, and small juvenile face with proptosis.

Figure 2—The limbs are unequally long and thick, with small hands and flat feet. There is lumbar lordosis and disproportion between the size of the limbs and the trunk.

FIG. 3

FIG. 4 FIG. 5

Figure 3—The increased width of the limbs is because of thickening of the bones, despite muscle wasting. The skin is rough, thick and dry. Figures 4 and 5—The shafts of the left humerus and right radius and ulna showing the absence of the normal trabecular pattern and an increase in density and width. The changes are most marked in the centre of the diaphysis but they reach almost to the epiphyses. The surface of the shaft is uneven. Note the proliferation of the cortex at the expense of the medulla.

CLINICAL INVESTIGATIONS

The examination of the blood gave the following results: Red blood count—5,200,000 per cubic millimetre. Haemoglobin—16 grammes per cubic millimetre. Colour index—1. Packed cell volume—50 per cent. Mean corpuscular volume—9 to 6·1 cubic microns. White blood count—7,000 per cubic millimetre. Differential white cell count—normal. Erythrocyte sedimentation rate—26 millimetres in one hour. Wassermann reaction—negative. Serum protein 6·4 grammes per 100 millilitres with a relative increase in the alpha globulin fraction. Serum calcium 10·4 milligrams per 100 millilitres. Serum phosphorus 4·3 milligrams per 100 millilitres. Serum alkaline phosphatase 23 King-Armstrong units. Serum cholesterol 210 milligrams per 100 millilitres.

The cerebrospinal fluid was normal, including the Wassermann reaction. The liver function and iodine uptake tests were also normal and the patient was able to excrete a water load within four hours with a normal rate of dilution. The electro-encephalograph showed a slight increase in fast activity.

RADIOLOGICAL FINDINGS

Long bones—There was an absence of the normal trabecular pattern with an increase in density and width of the shaft, most marked in the central part of the diaphysis but extending almost to the epiphysis. The outer margin of the shaft was uneven. The medulla was almost completely replaced by cortical expansion. These appearances were seen in all the long bones (Figs. 4 to 8).

Hands and feet—There was an increase in density of the shafts of the metacarpals and metatarsals with little widening. Similar but less marked changes were present in the phalanges (Figs. 9 and 10).

Pelvis—Dense areas were present in the ilium and ischiopubic rami on both sides, particularly the right (Fig. 11).

Skull—The base and all the calvarial bones were grossly thickened and dense except in the region of suture lines. The mandible was also affected but the facial bones were normal (Fig. 12).

Vertebrae—The cervical and upper thoracic vertebral bodies were flat but no more dense than normal (Fig. 13).

Ribs—There was a slightly increased density (Fig. 14).

Clavicles—These were dense and thick.

Scapulae—There were some dense areas in the bodies.

Air encephalograph—The ventricular system was normal in size, shape and position.

DISCUSSION

It is rare to find any two patients with a syndrome who are exactly alike, and the variation may be such that one may appear to have some manifestations of a second syndrome. After reviewing the literature on Engelmann's disease, we found it difficult to draw a clear-cut distinction between typical and borderline cases, particularly without personal knowledge of the patients concerned. Thus Paul (1953) compared his patients with those of Fritsch (1933), Ribbing (1949) and Feddema (Ribbing 1949) and observed that they showed features different from those of Engelmann's disease, because the latter had more widespread and symmetrical lesions, which had appeared at an earlier age and which were associated with disturbances of gait and posture. Later, however, Paul revised his opinion when he described the infant son of his first patient, who had all the typical features of Engelmann's disease.

Buchem *et al.* (1955) considered that their patients resembled those with Engelmann's disease, but did not feel that they should be classified as such. These patients, however, so closely resemble the patient described here that it seems certain that they should be included in

FIG. 6

FIGS. 6 TO 8

Figure 6—The left femur shows the same changes as those in the other long bones.

Figure 7—The lower ends of right and left femora and upper ends of both tibiae and fibulae show that the bone changes extend almost to the epiphyses.

Figure 8—The right and left tibiae and fibulae show the same changes as the other long bones.

FIG. 7

FIG. 8

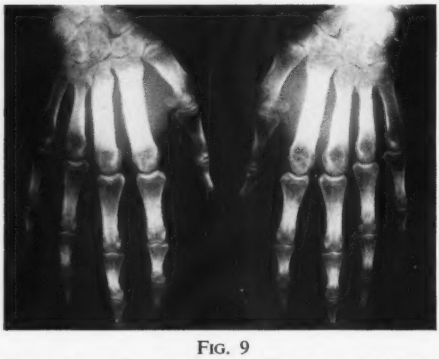


FIG. 9

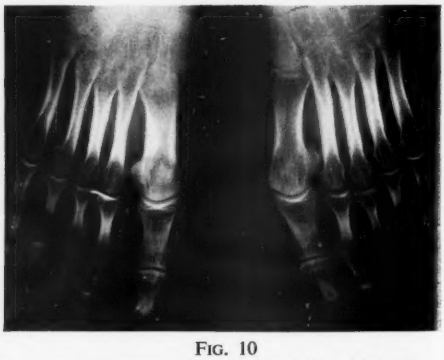


FIG. 10

The radiographs of the hands (Fig. 9) and feet (Fig. 10) reveal an increase in the density of the mid-shafts o the metacarpals, metatarsals and phalanges, particularly in the second, third and fourth rays.

this syndrome. A condition which the authors consider to be different from Engelmann's disease is leontiasis ossea with generalised bone changes as reported by Garland (1946) and Pygott and Scott (1954). We could find no patient with generalised bone sclerosis among the patients with leontiasis ossea described by Knaggs (1926).

FIG. 11

The radiograph of the pelvis shows dense areas in the ilium and ischiopubic rami on both sides but more on the right. The femoral heads are flattened and there is a valgus deformity of both femoral necks.

FIG. 12

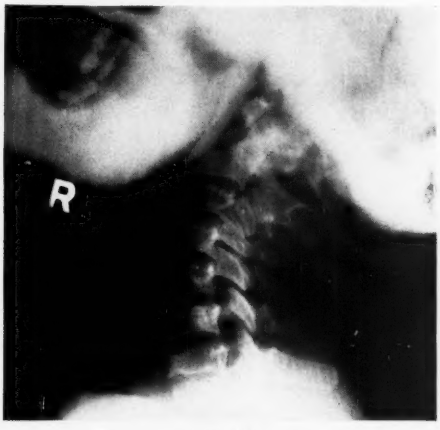


FIG. 13

Figure 12—A radiograph of the skull shows that the base and all the calvarial bones are thickened with sparing of bone adjacent to the sutures. There is enormous thickening of the frontal bone and the mandible is also affected.
Figure 13—Radiograph of cervical vertebrae. Note the flattening of the bodies of the vertebrae, without increase in density. Note the dense mandible and marked dental caries.

The condition known as generalised hyperostosis with pachydermia or pachydermoperiostitis (idiopathic osteoarthropathy) resembles hypertrophic osteoarthropathy. It begins at puberty with intermittent swelling of the hands and feet, increased width and length of the limbs, and is sometimes associated with pachydermia of the forehead and scalp. Radiologically

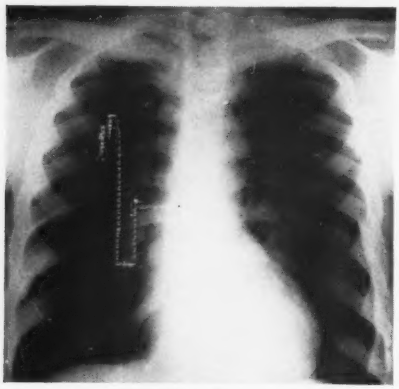


FIG. 14

Radiograph of chest showing slight increase in density and thickening of the ribs. The clavicles are affected and there are some dense areas present in the bodies of the scapulae.

the appearance of the bones resembles that of the most severe forms of Engelmann's disease except that there is a heavy, widely spaced trabecular pattern, rather different from the amorphous appearance in Engelmann's disease.

This discussion embraces fifty-two patients whom the authors consider to have had Engelmann's disease, including our own patient. The patients of Cockayne (1920), Ribbing (1949), Fritsch (1933), Feddema (Ribbing 1949), Lauterburg (1931), Buchem *et al.* (1955) and Farreras Valenti, Vilaseca and de Caralt (1954) have been included, but not those with leontiasis ossea with generalised bone changes or those with hyperostosis generalisata with pachydermia, although these conditions do possibly bear some relation to Engelmann's disease. Griffiths (1956) and other authors were of the opinion that Ribbing's report of his own and Feddema's patients had another disease, but when they are compared with those of Paul (1953) there seems to be little difference.

Age—The age was not stated in two patients. Of the remaining fifty the age at which symptoms started is shown in Table I. The youngest patient was three months old, and the oldest fifty-six years.

TABLE I

AGE AT WHICH THE SYMPTOMS OF ENGELMANN'S DISEASE STARTED

Age in years . .	0–10	11–20	21–30	31–40	41–50	51–60
Number of patients	19	8	11	3	4	5

Sex—There were thirty-one men and twenty-one women.

Race—Reports come from many parts of Europe, the United States of America, Canada and Australia. Wiedemann's (1948) patient was said to be of mixed German, Spanish and Negro descent, and there have been patients from an Iraqui family (Roth 1957) and a Filipino (Gulledge and White 1951). Lavine and Koven's (1952) patient was a Jew.

Heredity—It is likely that a genetic trait exists, for there are nine examples of familial incidence: father and son (Camurati 1922), four siblings (Ribbing 1949), five siblings (Feddema, cited by Ribbing), father and son (Lauterburg 1931), two siblings and a son (Paul 1953), two siblings (Ortolani and Castagnari 1953) and a father and two sons (Roth 1957).

Symptoms and signs—Pains in the limbs were present in twenty-three patients; described as aching or stabbing in nature and usually intermittent, the pain was worse after exercise and sometimes worse at night. A common site for the pain was one or both legs below the knee.

Being easily tired, delay in starting to walk, and being unable to run about are probably related to muscular weakness, which may be also responsible for the abnormal gait (twenty-five patients), lumbar lordosis (ten patients), flat feet (ten patients) and valgus deformity of the ankles (four patients). Thin legs or poor musculature were found in twenty-two patients, and an interesting observation was that in twelve patients the limbs seemed disproportionately long in comparison with the trunk and in two patients the bones were of unequal length. Failure to thrive, retarded growth or general underdevelopment was seen in thirteen patients. Signs of delayed puberty, hypogonadism, diminution of secondary sex characteristics, absence of pubic, axillary or facial hair were found in eleven patients.

A dry hard skin has been mentioned in five patients and absence of the subcutaneous fat layer in three.

Gross dental caries was a common finding. Four patients had a history of delayed dentition and in three the teeth were said to be widely spaced.

The presence of proptosis in six patients can be explained by diminution in the size of the orbital cavity from expansion of the surrounding bone; a decrease in the size of the cranial cavity and in the exit foramina of the eighth nerve probably accounted for the headache and deafness in four patients. Tendon reflexes in the affected limbs have usually been hyperactive but in four patients were hypoactive. Engelmann's own patient and four others were mentally dull.

Four patients had enlarged spleens and seven were anaemic.

An arbitrary division of patients into three groups can be made according to the severity of the disease.

Severe type—These usually have the typical posture and gait and a history from infancy of leg pains and difficulty in walking with or without muscle weakness. Most are diagnosed before puberty, but Stronge and McDowell's (1950) patient was twenty-eight, Michaelis's (1949) was twenty-four, and our patient was thirty-four.

Moderate type—These patients have either a history of past symptoms—such as the father described by Camurati (1922)—or a history of recent onset. Some have the typical posture and gait, but others are of normal physique. The bone changes are usually confined to a short length of the diaphysis in a few bones only, but they may be more extensive and out of proportion to the moderate severity of the clinical findings.

Mild type—These patients are asymptomatic and have been diagnosed radiologically on investigation of the families of patients belonging to the severe or moderate group. Examples are found in the reports of Ribbing (1949) and Paul (1953).

A family that has two or more of its members belonging to different groups suggests that these patients are suffering from different degrees or stages of the same condition.

Radiological changes—At any particular age the radiological changes vary with the severity of the clinical picture and are more marked in patients with a long history and in those with muscular weakness.

Usually the radiological progress is slow, but in Weingraber's (1954) case it was rapid. The changes start at any age and may become stationary. Bone has been seen to re-form in six weeks after biopsy (Singleton *et al.* 1956).

At first there is an increased density and widening of the cortex of the tibial or the femoral diaphysis on one or both sides, with a slight fusiform enlargement, but a smooth surface. These changes may start anywhere in the diaphysis but usually in the middle third.

Gradually the affected area involves the shaft towards the metaphyses and other bones may become affected. Later there is considerable encroachment on the medullary cavity, the width of the shaft increases, and the surface of the bone becomes irregular.

Finally the medullary cavity becomes almost obliterated and the whole bone, with the exception of the epiphyses, has a dense, amorphous appearance, less severe near the medullary cavity and epiphysis. The surface of the bone is now very irregular, but, because of increase in width of the whole shaft, the diameter of the bone is almost uniform throughout its length.

THE AFFECTED BONES

In the long bones the changes are usually bilateral and symmetrical. Two of Feddema's patients (Ribbing 1949) and one of Ribbing's (1949) had isolated bones affected as did Fritsch's (1933) patient when first seen. Unilateral involvement of one bone with bilateral affection of other bones has also occurred (Figs. 15 and 16).

Involvement of the tibia was most common, being present in forty-seven patients. The femur, fibula, humerus, radius and ulna followed in that order of frequency.

The base of the skull was affected in twenty-seven patients, usually in the anterior and middle fossae. In the vault the frontal bone was most frequently involved, usually with the density extending upwards from the orbital plates. No calvarial bone is exempt, and the facial bones have been affected in four patients. Narrowing of the exit foramina of the cranial nerves caused neurological changes in four patients.

When the pelvis is involved it is usually in the ilium near the acetabulum and close to the sacro-iliac joints.

Increase in density of the vertebrae has been mentioned in five patients; in four of these it was only the cervical vertebrae, and in two only the atlas, or the atlas and axis, that were affected, but all the vertebrae were affected in the patient reported by Gvozdanović (1950).

Roth (1957) remarked on the flattened vertebral bodies in his first patient. In our patient lateral radiographs of the spine show that the antero-posterior dimension of the vertebral

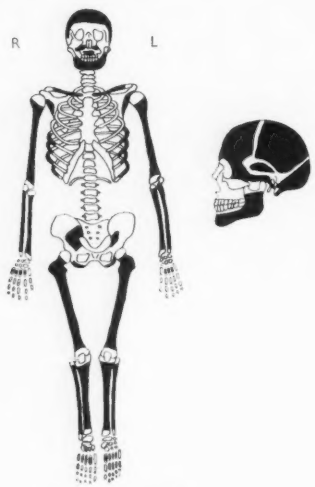


FIG. 15

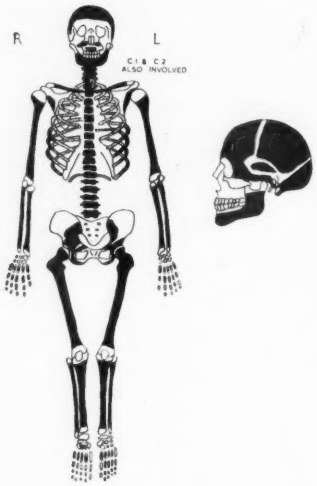


FIG. 16

Figure 15—Schematic representation of skeleton of the patient described in this report. The black shaded areas represent the bones involved and the extent of involvement. Figure 16—Schematic representation of skeleton, with blackened areas representing bones involved in reported cases in world literature. The dotted areas represent bones uncommonly affected.

bodies is increased relative to the height, so that the bodies do appear flattened, and this is particularly noticeable in the cervical spine. There is no increase in density; if anything, the bodies are slightly more translucent than normal.

The metatarsals and metacarpals have shown bilateral, but not always symmetrical, involvement in five patients, and the phalanges in two patients.

DIFFERENTIAL RADIOLOGICAL DIAGNOSIS

The similarities existing between Engelmann's disease, leontiasis ossea with generalised bone changes and hyperostosis generalisata with pachydermia have already been mentioned. Sear (1948) suggested that the well known—but uncommon—bone disorders, osteopetrosis and melorheostosis, may be related conditions.

When one bone is affected, before a diagnosis of Engelmann's disease can be made, Garre's osteomyelitis, Brodie's abscess, osteoid osteoma and neoplasm must be excluded.

When many bones are involved many diseases must be considered in the differential diagnosis, such as renal osteodystrophy, fibrous dysplasia, infantile cortical hyperostosis,

fluorosis, and heavy metal poisoning. A severe form of Engelmann's disease in the older age groups may resemble myelosclerosis, Paget's disease or osteoblastic carcinomatosis.

HISTOLOGICAL AND OTHER INVESTIGATIONS

There has been considerable variation in the histological findings, but this is to be expected when each specimen is of limited size, from different parts of various affected bones in different patients. In the radiologically altered bone there is increased cellularity with new bone formation. Fibrous tissue replacement of muscle fibres occurs. In the arteries proliferation of the intima and media has been seen, as well as in the veins of the subcutaneous tissues, muscle and bone marrow (Singleton *et al.* 1956, Neuhauser *et al.* 1948, Le Bien and Heilman 1951).

A multitude of laboratory investigations has usually revealed no abnormality. The patient of Farreras Valenti *et al.* (1954) had a positive Wassermann reaction.

PROGNOSIS

The only deaths recorded are those from anaemia at the age of three months (Gvozdanović 1950) and from pneumonia at fifty-two years (Buchem *et al.* 1955).

SUMMARY AND CONCLUSIONS

Engelmann's disease appears to be a familial and probably hereditary systemic condition, in which most of the manifestations can be attributed to a disorder in the osseous and muscular systems. At present the nature of the pathological process is obscure. The manifestations may present soon after birth or become apparent later in life. There is evidence that it is progressive but it may become arrested in some patients.

In order that this very incomplete state of knowledge may be improved it will be necessary to observe patients and their relatives throughout their lifetime.

We would like to thank Dr Hugh Davies for his valuable advice and help and Dr S. P. Meadows for permission to publish the case. Our thanks are also due to the Medical Committees of the National Hospital for Nervous Diseases and of the Hospital for Sick Children for allowing us access to their archives. We would also like to thank Mr A. H. Prickett of the Photographic Department of the Institute of Neurology for the photographs, and the secretarial staff of the Radiographic Department for preparing the manuscript.

REFERENCES

ANDERSON, F. G. (1953): Engelmann's Disease. *British Journal of Radiology*, **N.S. 26,** 603.

ANGEL, J. H. (1957): Pachydermo-periostosis (Idiopathic Osteoarthropathy). *British Medical Journal*, **ii,** 789.

BINGOLD, A. G. (1950): " Engelmann's Disease," Osteopathia Hyperostotica (Sclerotisans) Multiplex Infantilis; Progressive Diaphysial Dysplasia. *British Journal of Surgery*, **37,** 266.

BUCHEM, F. S. P. van, HADDERS, H. N., and UBBENS, R. (1955): An Uncommon Familial Systemic Disease of the Skeleton: Hyperostosis Corticalis Generalisata Familiaris. *Acta Radiologica*, **44,** 109.

CAFFEY, J. (1956): *Pediatric X-ray Diagnosis*. Third edition, p. 817. Chicago: The Year Book Publishers Inc.

CAMURATI, M. (1922): Di un raro caso di osteite simmetrica ereditaria degli arti inferiori. *Chirurgia degli Organi di Movimento*, **6,** 662.

CASUCCIO, C. (1949): *Osteopatie rare*, p. 432. Bologna: Edizioni Scientifiche Istituto Rizzoli.

COCCHI, U. (1951): Hereditary Diseases with Bone Changes. In *Roentgen Diagnostics*. By H. R. Schinz, W. E. Baensch, E. Friedl and E. Uehlinger. Vol. 1, Part 1, Section K, p. 702. London: William Heinemann Medical Books Limited.

COCKAYNE, E. A. (1920): Case for Diagnosis. *Proceedings of the Royal Society of Medicine* (Section for the Study of Disease in Children), **13,** 132.

COOLBAUGH, C. C. (1952): Effects of Reduced Blood Supply on Bone. *American Journal of Physiology*, **169,** 26. (Cited by Singleton.)

ENGELMANN, G. (1929): Ein Fall von Osteopathia hyperostotica (sclerotisans) multiplex infantilis. *Fortschritte auf dem Gebiete der Röntgenstrahlen*, **39,** 1,101.

FAIRBANK, Sir T. (1951): *An Atlas of General Affections of the Skeleton*. Edinburgh and London: E. & S. Livingstone Ltd.

FARRERAS VALENTI, P., VILASECA, J. M., and DE CARALT, M. (1954): Osteosclerosis diafisaria múltiple hereditaria tipo Camurati-Engelmann con síndrome de leontiasis ósea. *Revista Española de Reumatismo*, **5,** 354.

FEDDEMA, cited by RIBBING, S. (1949).

FELD, H., SWITZER, R. A., DEXTER, M. W., and LANGER, E. M. (1955): Familial Metaphyseal Dysplasia. *Radiology*, **65,** 207.

FRITSCH, H. (1933): Ein Fall von generalisierter Osteosklerose. *Wiener Archiv für Innere Medizin*, **23,** 247.

GARLAND, L. H. (1946): Generalized Leontiasis Ossea. *American Journal of Roentgenology*, **55,** 37.

GESCHICKTER, C. F., and COPELAND, M. M. (1949): *Tumors of Bone.* Third edition, p. 765. Philadelphia, London, Montreal: J. B. Lippincott Company.

GILLESPIE, J. B., and MUSSEY, R. D. (1951): Progressive Diaphyseal Dysplasia (Engelmann's Disease). *Journa of Pediatrics*, **38,** 55.

GRIFFITHS, D. Ll. (1956): Engelmann's Disease. *Journal of Bone and Joint Surgery*, **38–B,** 312.

GULLEDGE, H. W., and WHITE, J. W. (1951): Engelmann's Disease (Progressive Diaphyseal Hyperostosis) Report of a Case. *Journal of Bone and Joint Surgery*, **33–A,** 793.

GVOZDANOVIĆ, V. (1950): Ein neuer Fall von Engelmannscher Krankheit. Beitrag zur Kenntnis der kongenitale Osteodystrophien. *Fortschritte auf dem Gebiete der Röntgenstrahlen*, **73,** 86.

JACKSON, W. P. U., HANELIN, J., and ALBRIGHT, F. (1954): Progressive Diaphyseal Dysplasia. *Archives o Internal Medicine*, **94,** 902.

JAMMES, A., SERUZ, R., PROUXET, J., and DUCLOS, G. (1953): Maladie d'Engelmann: ostéopathie hyperostosant et sclérosante infantile multiple. (A propos d'une nouvelle observation.) *Revue du Rhumatisme*, **20,** 406.

KNAGGS, R. L. (1926): *The Inflammatory and Toxic Diseases of Bone.* Bristol: John Wright and Sons Ltd.

KOMINS, C. (1954): Familial Metaphyseal Dysplasia (Pyle's Disease). *British Journal of Radiology*, **N.S. 27,** 670.

KROSCH, H. (1955): Ein Fall von periostaler Hyperostose. *Fortschritte auf dem Gebiete der Röntgenstrahlen*, **83,** 546.

LAUTERBURG, W. (1931): Über 2 Fälle von familiärer generalisierter Osteosklerose. *Deutsche Zeitschrift für Chirurgie*, **230,** 308.

LAVINE, L. S., and KOVEN, M. T. (1952): Engelmann's Disease (Progressive Diaphyseal Dysplasia). *Journal of Pediatrics*, **40,** 235.

LE BIEN, W. E., and HEILMAN, C. (1951): Progressive Diaphyseal Dysplasia with Report of a Case. *Journal-Lancet*, **71,** 189.

MICHAELIS, L. S. (1949): Engelmann's Disease. *Proceedings of the Royal Society of Medicine* (Section of Orthopaedics), **42,** 271.

NEUHAUSER, E. B. D., SHWACHMAN, H., WITTENBORG, M., and COHEN, J. (1948): Progressive Diaphyseal Dysplasia. *Radiology*, **51,** 11.

ORTOLANI, M., and CASTAGNARI, G. (1953): L'osteopatia di Camurati-Engelmann. *Archivio "Putti" di Chirurgia degli Organi di Movimento*, **3,** 146. (Cited by Griffiths.)

PAUL, L. W. (1953): Hereditary Multiple Diaphyseal Sclerosis. (Ribbing.) Radiology, **60,** 412.

PERASSI, F. (1954): La malattia di Camurati-Engelmann. Iperostosi sclerotica diafisaria simmetrica ereditaria. *Radiologia Medica*, **40,** 147.

PYGOTT, F., and GILBERT SCOTT, M. (1954): Leontiasis Ossea (Virchow Type) with Report of a Case Showing Generalised Bone Changes. *British Journal of Radiology*, **N.S. 27,** 31.

RIBBING, S. (1949): Hereditary, Multiple, Diaphyseal Sclerosis. *Acta Radiologica*, **31,** 522.

RILEY, C. M., and SCHWACHMAN, H. (1943): Unusual Osseous Disease with Neurologic Changes. Report of Two Cases. *American Journal of Diseases of Children*, **66,** 150.

ROTH, J. (1957): Hyperostosis Generalisata, Camurati-Engelmann Type. *Harefuah*, **52,** 229.

SEAR, H. R. (1948): Engelmann's Disease. Osteopathia Hyperostotica Sclerotisans Multiplex Infantilis. Report of a Case. *British Journal of Radiology*, **N.S. 21,** 236.

SINGLETON, E. B., THOMAS, J. R., WORTHINGTON, W. W., and HILD, J. R. (1956): Progressive Diaphyseal Dysplasia. (Engelmann's Disease.) *Radiology*, **67,** 233.

STRONGE, R. F., and MCDOWELL, H. B. (1950): A Case of Engelmann's Disease. Progressive Diaphysial Dysplasia. *Journal of Bone and Joint Surgery*, **32–B,** 38.

THIERSCH, J. B. (1948): Contribution to the Radiology and Pathology of Transmissible Avian Osteopetrosis-Lymphomatosis. *Radiology*, **51,** 100.

UEHLINGER, E. (1943): Hyperostosis generalisata mit Pachydermie. *Fortschritte auf dem Gebiete de Röntgenstrahlen*, **67,** 8.

WEINGRABER, H. (1954): Eine neue Beobachtung bein einem Fall von osteopathia hyperostotica Engelmann-Camurati. *Fortschritte auf dem Gebiete der Röntgenstrahlen*, **81,** 800.

WIEDEMANN, H. R. (1948): Systematisierte sklerotische Hyperostose des Kindesalters mit Myopathie. *Zeitschri für Kinderheilkunde*, **65,** 346.

SCOLIOSIS IN BINOVULAR TWINS

BEN L. HULL, ALTOONA, PENNSYLVANIA, UNITED STATES OF AMERICA

From the Gorgas Hospital, Ancón, Panama Canal Zone

Murdoch (1959) reviewed the literature of idiopathic scoliosis in identical twins and reported a pair who had developed idiopathic scoliosis almost identical in site and at about the same age. He suggested that, since this condition seemed to depend on hereditary factors, there was a present need for reporting all cases of twins with scoliosis, including the occurrence in one or both members of a pair of binovular twins. In this paper I present examples of idiopathic scoliosis in binovular twins.

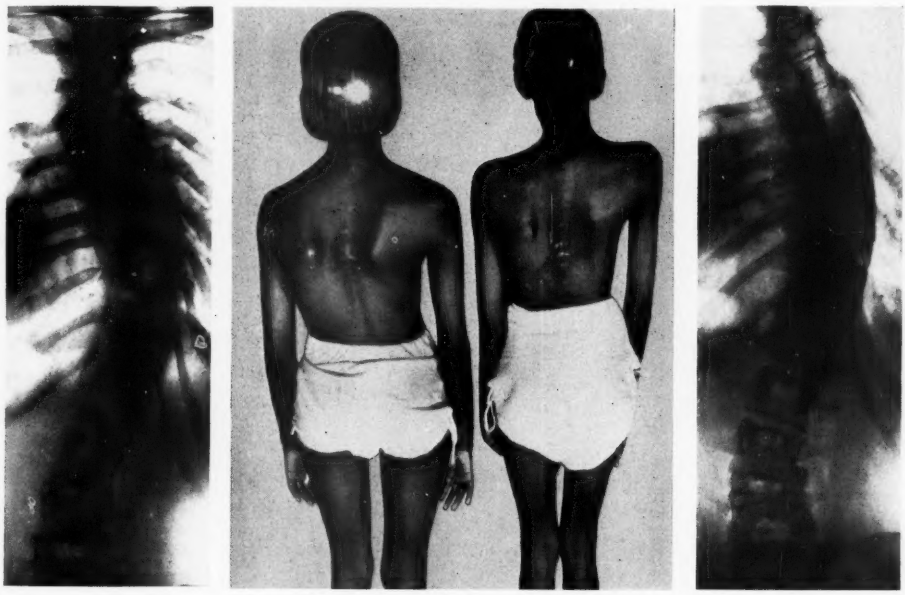


FIG. 1

Radiographs and photographs showing the similarity of the curves.

These children presented themselves at the age of fifteen. The deformity was a little more marked in one than in the other and more of the razor-back type deformity was present. There are seven children in this family, including another pair of binovular twin boys. There is no evidence of scoliosis in any other member of the family or in the parents.

It is hoped this report will stimulate others to report their cases. Eventually it might be well to have a scoliosis register on the lines of the bone tumour registries.

REFERENCE

MURDOCH, G. (1959): Scoliosis in Twins. *Journal of Bone and Joint Surgery*, **41–B,** 736.

HETEROTOPIC OSSIFICATION IN PARAPLEGIA

A Clinical Study

M. DAMANSKI, SOUTHPORT, ENGLAND

From the Liverpool Regional Paraplegic Centre, Promenade Hospital, Southport

This paper is based on study over twelve and a half years of 170 patients suffering from traumatic paraplegia or tetraplegia, of whom 162 survived for more than a month after injury and of 103 patients with other lesions of the spinal cord. The patients in the second group were less suitable for analysis because in many the lesions were disseminated or incomplete few could be compared with the patients with traumatic paraplegia, who generally had transverse and complete lesions with a well defined neurological level. Heterotopic ossification was found in forty-seven patients with traumatic paraplegia and twenty-five with non-traumatic lesions.

HISTORY

Towards the end of the first world war Déjerine and Ceillier presented to the Société de Neurologie in Paris a series of cases of heterotopic ossification in paraplegia, and published a comprehensive paper (1918c) containing a clinical and radiological study of the condition as observed at the Institution Nationale des Invalides. The following year Déjerine, Ceillier and Déjerine (1919) presented a paper on the anatomical and histological aspects of the disease, presenting sixteen anatomical specimens and a number of histological slides and colour photographs obtained from eleven patients. Little was known about the condition before these studies. After a prolonged search of the French literature for previously published cases of arthropathy in paraplegia, they were able to find only one unequivocal example (Chipault 1900). Geldmacher (1925) was unable to find records of more than four examples in the German literature (Riedel 1883, Israel 1920).

In the following years the condition was noted by other authors in traumatic and non-traumatic lesions of the central nervous system but was usually referred to as myositis ossificans. Geldmacher (1925) proposed the name ostiasis neurotica para-articularis or paraossalis. Soule (1945) called the condition neurogenic ossifying fibromyopathy. Nevertheless the term ectopic bone or heterotopic ossification seems the most appropriate designation because it covers bone formed in muscle and in other soft tissues, whether of inflammatory or non-inflammatory origin.

MORBID ANATOMY

Little can be added to Déjerine and her colleagues' (1918a, b, c; 1919) description of the morphology of heterotopic ossification in paraplegia. The main feature is the formation of new bone in the vicinity of an *unchanged* hip or knee joint. Thus the pathological characteristics are completely different from those of osteoarthritis and of the arthropathy of an unprotected joint (Charcot 1868).

They divided their patients into two main groups: those showing the heterotopic formation of bone in close relation to joints and bones, and those in whom the bone formed under the aponeurosis of or inside a muscle and had no connection with the skeleton. Such heterotopic bone was in the form of long needles or blades. The hip was the most common site for para-articular formation of bone. Sometimes the joint became fixed by the completion of a bony arch growing between femur and the innominate bone. Parosteal formation of bone most commonly started in relation to the medial femoral condyle and progressed upward in the soft tissues on the medial side of the bone. The lower part of the heterotopic bone was compact and the upper part spongy. The whole mass was joined to the femur by two or three projections. Heterotopic ossification was not found below the knee or above the level of paralysis.

HISTOLOGY

Heterotopic ossification consists of modified true bone, complete with Haversian canals, osteoblasts, blood vessels and oedematous bone marrow; in some instances there are also inclusions of cartilaginous tissue. Déjerine and her colleagues (1919) found no evidence of old or fresh extravasated blood or of pus. Where there is contact with the muscle, heterotopic ossification is separated from it by fibrous tissue, often dissociating the fascicles of the muscle. There is a perimuscular sclerosis often with atrophy of the muscle.

Déjerine (*loc. cit.*) did not find heterotopic ossification in the vicinity of pressure sores. However, Lhermitte (1919) found, on post-mortem examination, collections of pus and colonies of streptococci in one of his patients affected by bilateral and symmetrical heterotopic ossification.

Déjerine and Ceillier (1918c) did not observe heterotopic ossification in patients with neurologically incomplete lesions.

THEORIES OF PATHOGENESIS

Various theories regarding the formation of new bone in soft tissues in paraplegia have been put forward.

Déjerine and Ceillier (1918c) thought that there were two factors responsible for the formation of heterotopic ossification—a local one, and a central factor of neurogenic origin. The essential local factor was thought to be the lowering of the resistance of connective tissues by persistent oedema. Their failure to find heterotopic ossification in patients with incomplete lesions and their observation that heterotopic ossification was most frequent and extensive in patients with lesions sparing the intermedio-lateral column of the grey matter led them to the idea of a central, neurogenic, cause. They thought that toxaemia caused persistent irritability of the cells in this column which led to autonomic effects such as oedema hyperidrosis and muscular fibrillation. The combination of the two factors was thought to cause heterotopic formation of bone. Charcot (1877) before them had blamed the anterior horn cell for the same influence.

Léri (1919), applying Wolff's theory, believed that pressure and traction resulted in the formation of the tissue—bone—which was best able to meet the altered requirements. Lhermitte (1919) added a belief in the necessity of a superimposed factor of local infection. Israel (1920) put forward a similar theory of exaggerated reaction to pull and pressure.

Leriche and Policard (1918, 1926) suggested that heterotopic ossification in general develops in an ossifiable connective tissue near a depot of calcareous salts. This theory is based on Reichert's (1845) concept of the non-specificity of the various types of connective tissue derived from the mesenchyme. Leriche and Policard thought of the transformation of mature connective tissue to bone as taking place in three stages: 1) the mature connective tissue reverted to a primitive state as a result of infection or trauma; 2) the connective tissue became infiltrated with a collagenous hyaline substance arranged in lamellae, to form osteoid tissue; 3) this pre-osseous substance was converted to bone by impregnation with calcareous salts. This last process was only possible in the area of an excessive concentration of calcareous matter, released, in their view, by hyperaemia and decalcification of the adjacent bone.

Several hypotheses have been put forward to explain the local precipitation of calcium and phosphorus. In the opinion of Armstrong-Ressy, Weiss and Ebel (1957), the reduction in the serum protein concentration due to marked negative protein balance following spinal cord injury, together with the lowered carbon dioxide tension caused by the lowered metabolism in the paralysed muscles tend to favour the deposition of calcium salts. The role of phosphatase (Robison and Soames 1924) has also been extensively discussed.

CLINICAL FEATURES

To establish exactly the natural history of heterotopic formation of bone it would be desirable to examine the potential areas systematically from the very onset of paraplegia;

this has not been done in all the patients in this series. In this Centre no heterotopic ossification was recorded during the first month after injury to the spine.

The hips and knees are the areas of predilection, the ischial tuberosity and the upper limbs in tetraplegics being less frequently affected. The changes may be unilateral or bilateral and are often multiple. Clinical signs are present only when the condition is advanced. In most cases the condition was discovered as an incidental finding when radiographs were taken for other conditions.

Thickening around the hips and knees may be seen and felt in advanced cases, most commonly after prolonged nursing in the prone position. There may be limitation of flexion of the hips and knees, and in fewer cases limitation of extension; the stiffening of the hips may be associated with a lateral deviation of both lower limbs to one side. In the region of the knee the mass of the bone is fixed; there may be a fair range of movement, pain being elicited when the limb is being locked in a walking caliper.

In advanced cases of ankylosis in extension the patient finds it difficult to sit comfortably in his chair, although later he often learns to compensate for the fixed extension by placing his buttocks on the very front of the wheel-chair seat, his feet resting on the extended foot tray. Such a patient cannot use a toilet of the low-suite type, as the cistern is in his way.

This writer has not seen a subluxation or dislocation of the knee in these cases.

Heterotopic ossification of a special nature may be associated with the paralysis caused by extension injuries of the neck in elderly patients. Shortly after the spinal injury these patients often complain of pain in their shoulders, elbows and hands, and of swelling of the hands. If they are not given adequate physiotherapy they develop stiff joints and later ossification may occur in the adductors of the shoulder, in the cubital fossa, or in the triceps tendon, with a fixed contracture of the joints.

RADIOGRAPHIC APPEARANCES

Heterotopic ossification does not appear in a haphazard manner, but follows certain patterns which may be seen in combination. One of the main features is that the condition is extra-articular and extra-capsular.

Déjerine and Ceillier (1918c) pointed out that there was usually osteoporosis in the adjacent bones, as if the calcareous matter used for the formation of ectopic bone had been borrowed from the vicinity. In the writer's experience even a moderate degree of decalcification is uncommon, and a local decalcification was only observed in one case in the bones of the knee joint. Perhaps the technique of microradiography and autoradiography with radioactive tracers as reported by Lacroix (1960) will provide us with more accurate means of assessing the presence and degree of decalcification.

Ossification around the hip—On the femoral side the bone usually forms in relation to the great trochanter (Fig. 1), less commonly in relation to the trochanteric crest (Fig. 2) or lesser trochanter (Fig. 3).

On the pelvic side the bone may form on the lateral side of the junction of the body and ramus of the ischium, and may then be associated with irregularity of the ischium (Fig. 4). It may also form in relation to the anterior inferior spine of the ilium (Fig. 5).

There seems to be a tendency for the formations to grow together from each side. Sometimes they fuse to bridge the joint, most commonly above it (Fig. 6) but also below it (Fig. 7). A complete extra-articular ankylosis may result (Fig. 8). The joint itself remains unaffected unless involved in infection introduced through a pressure sore (Fig. 9).

The bone is connected with the skeleton to a variable extent. In some cases it may appear radiologically to be continuous with the bones of the hip joint; in others it is clearly seen to be separate from them. In the latter group, most formations of bone seem to be in the gluteus medius, while others seem to be in the adductor muscles and the pectineus or in the quadriceps. Occasionally, bone seems to form in the tensor fasciae latae. In one case bone of a perforated

FIG. 1

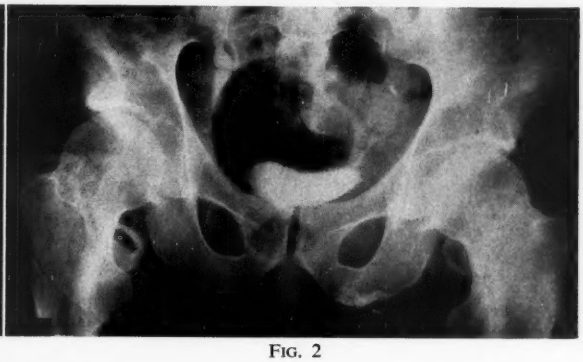


FIG. 2

FIG. 3

FIGS. 1 TO 3

Figure 1—Case 1. Man aged twenty-one with tetraplegia from transverse myelitis. Right trochanteric sore. Severe infection of urine; temporary loss of function of the left kidney. Note heterotopic formation of bone in relation to the great trochanter. Figure 2—Case 2. Man aged thirty-six with flaccid paraplegia below first lumbar segment from fracture-dislocation of twelfth thoracic on first lumbar vertebra. Bilateral trochanteric sores. Severe infection of urine; hydronephrosis. Nursed several months in prone position. Heterotopic bone arising from the trochanteric crests; needle-shaped heterotopic bone near the adductor origin from the ischium. Figure 3—Case 3. Man aged fifty-nine with spastic paraplegia below first lumbar segment due to vascular lesion of the cord. Left-sided trochanteric sore; patient bedridden in prone position for many months. Heterotopic ossification arising from the greater and lesser trochanters of the left femur; note the stalactite-stalagmite effect; contiguity but no continuity of the ectopic bone at the greater trochanter.

FIG. 4

FIG. 5

Figure 4—Case 4. Man aged fifty-four with flaccid paraplegia below eleventh thoracic segment from fracture-dislocation of eleventh on twelfth thoracic vertebra. Bilateral trochanteric sores; severe infection of urine; hydronephrosis; chronic uraemia. Patient nursed in prone position for many months. Heterotopic ossification at the junction of the body and ramus of the right ischium. Note contiguity, but no continuity; irregular outline of ischial tuberosity. Figure 5—Case 5. Man aged twenty-six with spastic paraplegia below sixth thoracic segment from extramedullary ependymoma. Right trochanteric sore; nursing in prone position. Heterotopic ossification arising from the outer aspect of the right ilium, at the level of the anterior inferior iliac spine.

FIG. 6

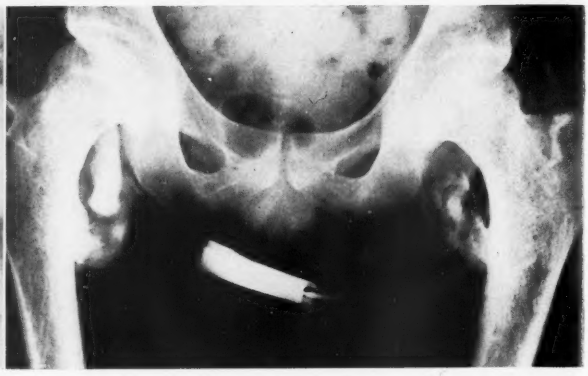


FIG. 7

Figure 6—Case 6. Man aged thirty-three with spastic paraplegia below ninth thoracic segment from fracture-dislocation of tenth on eleventh thoracic vertebra. Multiple pressure sores. Severe infection of urine. Prolonged nursing in prone position. Firm union between the right great trochanter and the outer ilium; bulky bony inclusions in the adductors; a small one in the right tensor fasciae latae. Figure 7—Case 7. Man aged thirty-seven with flaccid paraplegia below first lumbar segment from fracture-dislocation of twelfth thoracic on first lumbar vertebra. Ischial sore; prolonged nursing in prone position. Severe infection of urine; death from uraemia. Union between the lesser trochanters and the pelvis is taking place; slender bony needles near the adductor origins.

FIG. 8

FIG. 9

Figure 8—Case 8. Man aged twenty-one with spastic paraplegia below tenth thoracic segment from fracture-dislocation of ninth on tenth thoracic vertebra. Pressure sores over both greater trochanters. Severe urinary infection, chronic uraemia. Prolonged nursing in prone position. Complete bony ankylosis of the left hip in extension resulting from union in the upper and lower tier. Figure 9—Case 9. Man aged forty-two with flaccid paraplegia below tenth thoracic segment due to encephalo-myelitis. Dislocation of the right hip from septic arthritis caused through a trochanteric sore.

sponge-like appearance was seen to form in relation to the ischial tuberosities (Figs. 10 and 11). After removal of the ischium a horn-like bony proliferation may occur.

Ossification around the knee—The ligamentum patellae, either alone (Fig. 12) or with the quadriceps tendon (Fig. 13), is frequently the seat of ectopic bone.

Bone often forms in relation to the medial femoral condyle but much less commonly in relation to the lateral condyle and in that site to a much lesser extent (Fig. 14). Ectopic bone formation has been seen in relation to the tibial condyle (Fig. 15), but not below this level.

Ossification in the upper limb—Heterotopic ossification in the upper limb was seen in four patients aged between fifty and seventy, who had suffered extension injuries of osteoarthritic necks. It was not seen in patients paralysed as a result of flexion injuries.

FIG. 10

FIG. 11

Case 10—Figure 10. Man aged twenty-seven with flaccid paraplegia from trauma on quiescent Pott's disease. Deep pressure sore over the right ischial tuberosity. Spongy new bone along the distal border of the right ischium. Figure 11—Ischium removed at operation. Note the lace-like appearance of the heterotopic bone; the line of separation between the ischium and the new bone can easily be distinguished. (Scale in centimetres).

In one patient, a man of sixty-one, heterotopic bone formed bridges between the humeri and scapulae and also massively involved the anterior aspect of the left elbow (Figs. 16 and 17). In another patient formation of bone in the region of the attachment of the triceps was seen (Fig. 18).

ETIOLOGY OF HETEROTOPIC OSSIFICATION

It is possible to divide etiological factors into two main groups: those which remain beyond our control like the age of the patient and the level of the spinal injury, and those which depend on the type of treatment given.

Age—In this series the relative peak of spinal injury was in the fourth and third decades of life, whereas the relative peak of heterotopic ossification was in the seventh and sixth decades.

Level of spinal injury—In this series the most common level of injury was at the first lumbar vertebra. There was a gradual decline towards the twelfth and eleventh thoracic vertebrae, injuries at the thoraco-lumbar junction accounting for about 60 per cent of the cases. There was a lower peak of incidence at the cervical level, flexion and extension injuries together accounting for about 25 per cent. The lowest peak of incidence was at the thoracic region from the third to the tenth vertebrae, injuries in that region accounting for about 20 per cent of the total.

In contrast (Fig. 19), the peak incidence of heterotopic ossification was in patients with injuries at the eleventh thoracic vertebral level. The incidence diminished in the cervical and lumbar injuries, if extension injuries of the neck were excluded. In these, the incidence of heterotopic ossification was high.

FIG. 12 FIG. 13

Figure 12—Case 8. Man aged twenty-one with spastic paraplegia below tenth thoracic level from fracture-dislocation of ninth on tenth thoracic vertebra. Multiple pressure sores, but none over the patellae. Severe urinary infection, hydronephrosis, chronic uraemia. Nursed in prone position for several months. Substantial heterotopic ossification of the patellar tendon; also a spur in the quadriceps tendon. Figure 13—Case 11. Man aged fifty-four with flaccid paraplegia below eleventh thoracic segment from fracture-dislocation of eleventh on twelfth thoracic vertebra. Multiple pressure sores, but none over the affected knee. Severe urinary infection, chronic uraemia. Prolonged nursing in prone position. Ossification predominantly in the quadriceps tendon.

Neurological lesion—The relative incidence of heterotopic ossification in patients with fractures of the first lumbar vertebra, who usually showed a flaccid paralysis, was in proportion to the relative incidence of spinal fracture. There was a steep decline of incidence in the patients with flaccid paralysis due to injuries of the second to fifth lumbar vertebrae.

In the group with thoracic vertebral injuries the incidence of heterotopic ossification was highest in patients with injuries of the eleventh thoracic vertebra and with consequent spastic paralysis. The incidence declined in the patients with spastic paralysis produced by higher vertebral lesions. There was in general, little difference in the incidence of ossification between those with spastic and those with flaccid paralyses.

Only those patients with heterotopic ossification following cervical extension injuries had neurologically incomplete lesions.

FIG. 14

FIG. 15

Figure 14—Case 8. Man aged twenty-one with spastic paraplegia below tenth thoracic segment from fracture-dislocation of ninth on tenth thoracic vertebra. Severe infection of urine; hydronephrosis; chronic uraemia. Multiple pressure sores but none over the affected knee. Apart from a massive ossification near the medial condyle and a small one at the medial margin of the plateau of the tibia, there is another ossification near the lateral condyle with a diaphysial sponge-like extension. Figure 15—Case 12. Man aged forty-eight with spastic paraplegia below fifth thoracic segment from fracture-dislocation of fourth on fifth thoracic vertebra. Multiple pressure sores, but none over the affected knee. Severe urinary infection; hydronephrosis. Ossification below the plateau of the tibia.

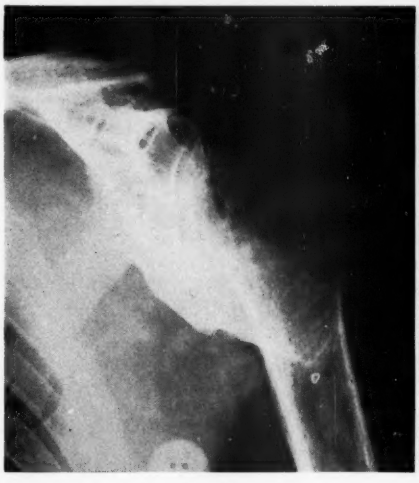


FIG. 16

FIG. 17

Case 13—Figure 16. Man aged sixty-one with incomplete tetraplegia from extension injury of the cervical spine. Inadequate physiotherapy in the early stage resulting in contracture in adduction of both shoulders and in flexion of the left elbow. Pressure sores; severe infection of urine; hydronephrosis. Note ossification in adductor muscles of shoulder. Figure 17—The left elbow. Note the ossification in the cubital fossa.

Management of spinal fracture—Most patients were admitted to the Centre some time after injury, their early treatment having been carried out elsewhere. The type of early treatment had some bearing on the incidence of heterotopic ossification (Table I).

Urinary infection and nephrolithiasis—In the present series the frequency of stone formation was about the same in those patients with heterotopic ossification and those without it. On the other hand, it was closely related to severe infection of the urinary system. It was also noted that the mortality from uraemia in the group with heterotopic ossification was much higher than in the other patients—26 as against 11 per cent.

FIG. 18

Case 14—Man aged fifty-two with incomplete tetraplegia from extension injury of the cervical spine. Flexion contractures of both elbows. Deep and extensive pressure sores which finally led to his death. Severe urinary infection. Note ossification in the triceps insertion.

Pressure sores—All the patients affected by heterotopic ossification had pressure sores.

Thus, ectopic bone at the ischial tuberosity was usually associated with a pressure sore on the adjacent skin. Similarly, sores were usually associated with heterotopic ossification in the coxo-femoral space. Four-fifths of the affected patients had sores on the ipsilateral or contralateral trochanteric areas, or both, and the remaining fifth in the other areas of the pelvic girdle, mainly over the sacrum or the buttocks. Heterotopic ossification in the region of the knee was rarely associated with a sore over the patella or one of the condyles.

TREATMENT

There is no effective treatment for heterotopic ossification. Excision of the bony mass has been tried (Miller and O'Neill 1949), also with reconstruction of the hip joint using a Judet prosthesis (Armstrong-Ressy *et al.* 1957). The operation very rarely led to a permanent arthrolytic effect only, owing to tendency to re-formation of heterotopic bone and to infection. In this Centre intertrochanteric osteotomy was performed in a surgically clean case and led to increased and quick formation of new bone around the site of operation and union of the fragments. No pseudarthrosis could be obtained.

TABLE I

INCIDENCE OF HETEROTOPIC OSSIFICATION RELATED TO TYPE OF PRIMARY TREATMENT

Method	Number of patients	Percentage affected by heterotopic ossification
Skull traction	12	0
Open reduction and internal fixation	15	20
Open reduction alone	7	29
Laminectomy	21	29
Plaster-of-Paris, either alone or following open reduction or laminectomy	24	46

Several cases of acute septic arthritis of the hip were seen resulting from trochanteric sores and associated with formation of ectopic bone. The routine treatment used in these

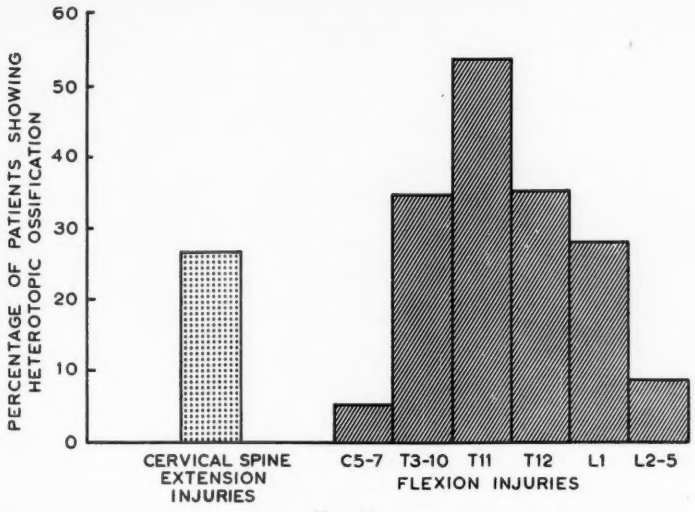


FIG. 19

Incidence of heterotopic ossification related to the bony level of injury. (Patients dying within the first month of injury have been excluded.)

patients at one time was resection of the femoral neck and head. This almost invariably led to ankylosis in extension because of exuberant heterotopic formation of bone. Nearly always sinuses persisted. In all these patients disarticulation of the lower limb through the hip was ultimately necessary; amputation was followed by exuberant formation of bone on the outer aspect of the ilium (Fig. 20).

Thus, amputation of the leg seems to be the only dependable operation in surgically clean cases when a patient suffers from intolerable discomfort or when there are persistent sinuses resulting from excision of the head of the femur. Very few patients indeed will need this mutilating procedure.

Fleming (1957) reported a doubtful clinical improvement after treating two patients with ultrasonic waves.

FIG. 20

Case 15.—Man aged thirty-three with spastic paraplegia below ninth thoracic segment from fracture-dislocation of tenth on eleventh thoracic vertebra. Pressure sores. Severe urinary infection. Heterotopic ossification at the outer aspect of the left ilium after disarticulation through the hip.

PROPHYLACTIC TREATMENT

It is the writer's experience that, with a few exceptions, heterotopic ossification in paraplegics results from failure to give these patients the best treatment from the first day of their disability. Severe concomitant injuries or an uncooperative attitude on the part of the patient may make ideal treatment impracticable. Heterotopic ossification tends to occur in association with other complications such as

TABLE II

INCIDENCE OF HETEROTOPIC OSSIFICATION RELATED TO TIME ELAPSING BETWEEN INJURY AND ADMISSION TO THE CENTRE

Year of admission	Number of patients	Heterotopic ossification		Number of patients admitted within fourteen days of spinal injury	Heterotopic ossification. (Patients admitted within fourteen days of injury)	
		Number of patients	Percentage		Number of patients	Percentage
1947	17	4	24	0	0	0
1948	5	3	60	0	0	0
1949	10	4	40	0	0	0
1950	13	6	46	0	0	0
1951	13	5	38	2	0	0
1952	12	7	58	4	0	0
1953	15	4	27	5	0	0
1954	17	6	35	8	3	37
1955	18	3	17	13	3	23
1956	7	1	14	4	1	25
1957	16	2	12	9	1	11
1958	13	2	15	6	0	0
1959	6	0	0	3	0	0
Total .	162	47	29	54	8	15

hypoproteinaemia, anaemia, pressure sores and severe infection of the urinary bladder. Under ideal conditions the risk of heterotopic ossification may be substantially reduced, or even eliminated altogether (Table II).

Déjerine and Ceillier (1918c) in their study of seventy-eight paraplegics found thirty-eight to be affected by heterotopic ossification, an incidence of 48 per cent. The average incidence in the present series was 29 per cent. The incidence for 1948 was 60 per cent, but since then the incidence has been declining step by step with improving standards of treatment.

In 1951 this Centre began to admit paraplegics shortly after injury and it was hoped that none of these " fresh " cases would develop heterotopic ossification, provided the various predisposing factors were avoided. In the main this hope was fulfilled.

In 1954 there were three failures: two elderly men with extension injuries of the cervical spine developed heterotopic ossification in the upper limbs; in retrospect it is felt that they had not received as much physiotherapy as was necessary. One patient had on admission pressure sores and severe infection of the bladder. In 1955 there were again three cases: one patient had on admission pressure sores and a badly infected bladder, and two patients who had been discharged with a poorly controlled urinary infection were found on readmission to have pressure sores, hydronephrosis and heterotopic ossification. Finally, in 1957, a patient with cervical extension injury developed heterotopic ossification in the biceps; once more it seemed that this was due to inadequate physiotherapy.

In this Centre the following prophylactic measures have been applied with success.

Maintenance of good general condition and avoidance of pressure sores—It has been known since the works of Cuthbertson (1930, 1932, 1936, 1942, 1954, 1957) that there is often an excessive excretion of nitrogen in the urine during the period immediately following a fracture

This in turn results in depletion in the body proteins. This occurs in most spinal fractures and the condition is made worse by severe concomitant injuries and early operative procedures. Diminished resistance of the skin to pressure and of the bladder to infection results. Pressure sores and urinary infection develop and lead to further loss of protein. Hypoproteinaemia leads also to oedema of the tissues. The negative nitrogen balance can be corrected to a certain extent by a high-protein diet but unfortunately most paraplegic patients display a considerable loss of appetite in the early stage of their illness and develop hypoproteinaemia with anaemia often lasting for many months. The writer considers that the most reliable method for avoiding and correcting the negative nitrogen balance is blood transfusion. Transfusion may have to be repeated frequently over a long period of time.

In the writer's opinion, oedema from hypoproteinaemia plays a considerable role in the pathogenesis of heterotopic ossification. Charcot (1868) and Déjerine and Ceillier (1918c) described oedema as a constant feature of paraplegia and in all probability what they were describing was a typical hypoproteinaemic oedema.

Constant change of position—Changing the patient's position in bed every two to four hours is fundamental to the treatment of paraplegia but, unfortunately, the principle had sometimes to be discarded in favour of a permanent or semi-permanent prone positioning of the patient in the event of pressure sores or thin scars over the sacrum, etc.

This is an unnatural position which involves shortening of the hip and knee extensors and stretching of the flexors, uncontrolled weight bearing by the anterior thigh muscles with a probable compression of the blood and lymph vessels. Almost all the patients affected by heterotopic ossification were nursed for periods of variable length in the prone position which often resulted in oedema of local origin restricted to the lower abdomen, inguinal regions, genitalia and the upper thighs, the parts of the body left clear of the proximal and distal sectional mattresses. It is this writer's experience that this was often associated with heterotopic ossification.

Early passive and active mobilisation—Frequent passive movements of the paralysed limbs were recommended a long time ago and this is the most dependable method not only for avoiding capsular and pericapsular changes but also for preventing shortening of one muscle group and stretching of the opponents, leading to the necessity for forceful and painful manipulation. This applied equally to the upper limbs in incomplete tetraplegia resulting from extension injuries where considerable shortening of the shoulder adductors and elbow flexors may take place over a period of a few days.

The importance of early mobilisation in a wheel-chair cannot be stressed enough. In case of a complete neurological lesion when no recovery of the lost functions can be expected the patients should be allowed out of bed as soon as the pain at the site of the spinal fracture and the condition of concomitant injuries do not stand in the way (Kerr 1956). In the neurologically incomplete lesions mobilisation in a wheel-chair commences as soon as the danger of increase in the collapse of the crushed vertebral body and of dislocation is over.

Early treatment of excessive spasticity—In the event of excessive spasticity of the lower limbs leading to flexion contractures of the hips and knees, anterior root section, from L.1 to S.1 inclusive, was the method of choice if the general condition permitted. Subarachnoid injection of alcohol was reserved for the poor operative risks. Both procedures were applicable in neurologically complete lesions and in the incomplete ones when the remaining neurological functions were of no practical value. In cases where the spasticity of the thigh adductors was the troublesome factor, an intrapelvic obturator neurectomy was performed. Care was taken to operate as soon as the spasticity had reached its presumable peak as the failure to deal with it early often leads to fixed flexion contractures and heterotopic ossification.

Avoidance of severe infection of the bladder—Practically every patient suffering from a spinal injury with neurological involvement has to go through a period of catheter drainage. Sooner

or later this causes infection varying in degree from a slight bacteriuria to a severe pyuria It is necessary to do everything possible to prevent or limit this infection.

At the Southport Centre this has been successfully achieved by observing the following rules: 1) A " fresh " patient is admitted to a special post-operative ward where there is less chance of cross-infection and where he remains until the catheter is no longer required 2) A long, polythene Gibbon catheter, connected directly to a Lane's bottle, is used as a closed-drainage system (Gibbon 1958; Ross, Gibbon and Damanski 1957). This catheter has the advantage of being non-irritant owing to its small size and chemical properties and there is no glass connection at the outer end near the bladder which, with other types of catheter tends to facilitate cross-infection when the catheter is temporarily disconnected from the bottle. 3) Whatever the patient's method of voiding, either by abdominal straining or by reflex contraction of the detrusor of the bladder, it should be as complete as possible, with no or little residual urine. This is achieved spontaneously, at a certain stage of treatment, or by surgical procedures, mainly by bladder-neck resection (Ross 1951, 1956) or division of the external sphincter (Ross, Damanski and Gibbon 1957, 1958; Ross, Gibbon and Damanski 1957, 1958). The indications for these procedures are established after several urological examinations. In this connection cysto-urethrography is considered to be of great value (Damanski 1961; Damanski and Kerr 1957). 4) Periodical intravenous pyelography is the best method for assessing the effect of the infection on the upper tracts, and to find at the earliest possible moment hydronephrosis, ureteric reflux (Ross, Damanski and Gibbon 1960), loss of renal function or stones.

The writer believes that infected pressure sores and infection of the bladder are the main two factors in the pathogenesis of heterotopic ossification.

SUMMARY

1. A clinical study has been made of heterotopic ossification in 273 patients with paraplegia of traumatic and non-traumatic origin treated at the Liverpool Paraplegic Centre over a period of twelve and a half years.

2. The literature is reviewed and theories of etiology are discussed.

3. Etiological factors have been studied. Prominent among these is inadequacy of early treatment leading to urinary infection and to the formation of pressure sores.

4. It is concluded that there is no effective treatment for established heterotopic ossification.

5. The importance of prophylactic treatment is stressed. Special emphasis is placed on adequate primary treatment, correction of hypoproteinaemia and early mobilisation.

REFERENCES

ARMSTRONG-RESSY, C. T., WEISS, A. A., and EBEL, A. (1957): Results of Surgical Treatment of Extraosseous Ossifications in Spinal Cord Injuries. *Proceedings of Sixth Annual Clinical Paraplegia Conference, October 22–24, 1957, Veterans Administration Hospital, Richmond, Virginia, U.S.A.*, p. 12.

CHARCOT, J. M. (1868): Sur quelques arthropathies. *Archives de Physiologie*, **1**, 161. Quoted by Charcot (1877).

CHARCOT, J. M. (1877): *Lectures on the Diseases of the Nervous System*, pp. 92–102. English Translation by G. Sigerson. London: The New Sydenham Society.

CHIPAULT, A. (1900): Les arthropathies trophiques consécutives aux affections chirurgicales de la moelle. *Traité de Neurologie Chirurgicale*, pp. 76–113. Quoted by Déjerine and Ceillier (1918).

CUTHBERTSON, D. P. (1930): The Disturbance of Metabolism Produced by Bony and Non-bony Injury, with Notes on Certain Abnormal Conditions of Bone. *Biochemical Journal*, **24**, 1,244.

CUTHBERTSON, D. P. (1932): Observations on the Disturbance of Metabolism Produced by Injury to the Limbs. *Quarterly Journal of Medicine*, N.S. **1**, 233.

CUTHBERTSON, D. P. (1936): Further Observations on the Disturbance of Metabolism Caused by Injury, with Particular Reference to the Dietary Requirements of Fracture Cases. *British Journal of Surgery*, **23**, 505.

CUTHBERTSON, D. P. (1942): Post-shock Metabolic Response. (Arris and Gale Lecture to Royal College of Surgeons of England.) *Lancet*, **i**, 433.

CUTHBERTSON, D. P. (1954): Interrelationship of Metabolic Changes Consequent to Injury. *British Medical Bulletin*, **10**, 33.

CUTHBERTSON, D. P. (1957): The metabolic changes consequent on injury. *Zeitschrift für Tierernährung und Futtermittelkunde*, **12,** 259.

DAMANSKI, M. (1961): Cysto-Urethrography in Paraplegia: Its Practical Application. *British Journal of Urology*, **33,** 67.

DAMANSKI, M., and KERR, A. Sutcliffe (1957): The Value of Cysto-urethrography in Paraplegia. *The British Journal of Surgery*, **44,** 398.

DÉJERINE, Mme, and CEILLIER, A. (1918a): Trois cas d'ostéomes. Ossifications périostés juxta-musculaires et interfasciculaires. *Revue Neurologique*, **25,** 159, 207. Quoted by Déjerine and Ceillier (1918c).

DÉJERINE, Mme, and CEILLIER, A. (1918b): Les para-ostéo-arthropathies des paraplégiques. Société de Neurologie, séance de juin 1918. *Revue Neurologique*, **25,** 491. Quoted by Déjerine and Ceillier (1918c).

DÉJERINE, Mme, and CEILLIER, A. (1918c): Para-ostéo-arthropathies des paraplégiques par lésion médullaire (Étude clinique et radiographique). *Annales de Médecine*, **5,** 497.

DÉJERINE, Mme, CEILLIER, A., and DÉJERINE, Yv. (1919): Para-ostéo-arthropathies des paraplégiques par lésion médullaire. Étude anatomique et histologique. *Revue Neurologique*, **26,** 399.

FLEMING, W. C. (1957): Preliminary Observations on the Use of Ultrasonics in Treatment of Soft Tissue Calcification in Paraplegics. *Proceedings of 6th Annual Clinical Paraplegia Conference, October* 22–24, 1957, *Veterans Administration Hospital, Richmond, Virginia, U.S.A.*, p. 18.

GELDMACHER, M. (1925): Beitrag zu den parartikulären Verknöcherungen nach Querschnittsläsion des Rückenmarks. *Deutsche Zeitschrift für Chirurgie*, **191,** 180.

GIBBON, N. (1958): A New Type of Catheter for Urethral Drainage of the Bladder. *The British Journal of Urology*, **30,** 1.

ISRAEL, A. (1920): Über Myositis ossificans neurotica nach Schussverletzung des Rückenmarkes. *Fortschritte auf dem Gebiete der Röntgenstrahlen*, **27,** 365.

KERR, A. Sutcliffe (1956): Personal communication.

LACROIX, P. (1960): Microtechniques for Studying Bone. *British Medical Journal*, **i,** 342.

LÉRI, A. (1919): Discussion on Déjerine's *et al.* paper. *Revue Neurologique*, **26,** 461.

LERICHE, R., and POLICARD, A. (1918): Sur la production expérimentale d'os chez l'homme adulte en dehors de toute action ostéopériostique. *Bulletins et Mémoires de la Société de Chirurgie*, **44,** 1,265. Quoted by Leriche and Policard (1926).

LERICHE, R., and POLICARD, A. (1926): *Les problèmes de la physiologie normale et pathologique de l'os.* Paris: Masson & Cie.

LHERMITTE, J. (1919): Discussion on Déjerine's *et al.* paper. *Revue Neurologique*, **26,** 460.

MILLER, L. F., and O'NEILL, C. J. (1949): Myositis Ossificans in Paraplegics. *Journal of Bone and Joint Surgery*, **31–A,** 283.

REICHERT, C. B. (1845): *Bemerkungen zur vergleichenden Naturforschung im Allgemeinen und vergleichende Beobachtungen über das Bindegewebe und die verwandten Gebilde.* Dorpat: W. Gläser. Quoted by Leriche and Policard (1926): p. 11; and Brailsford, J. F. (1948): *The Radiology of Bones and Joints*, p. 591. London: J. & A. Churchill Ltd.

RIEDEL, B. (1883): Demonstration eines durch achttägiges Umhergehen total destruirten Kniegelenkes von einem Patienten mit Stichverletzung des Rückens. *Verhandlungen der Deutschen Gesellschaft für Chirurgie*, **12,** 93.

ROBISON, R., and SOAMES, K. M. (1924): The Possible Significance of Hexose-Phosphoric Esters in Ossification. Part II. The Phosphoric Esterase of Ossifying Cartilage. *Biochemical Journal*, **18,** 740. Quoted by Leriche and Policard (1926).

ROSS, J. Cosbie (1951): Management of the Bladder in Spinal Injuries. *British Medical Journal*, **i,** 616.

ROSS, J. Cosbie (1956): Treatment of the Bladder in Paraplegia. *British Journal of Urology*, **28,** 14.

ROSS, J. Cosbie, DAMANSKI, M., and GIBBON, N. (1957): Resection of the External Urethral Sphincter in the Paraplegic—Preliminary Report. *Transactions of the American Association of Genito-Urinary Surgeons*, **49,** 193.

ROSS, J. Cosbie, DAMANSKI, M., and GIBBON, N. (1958): Resection of the External Sphincter in the Paraplegic—Preliminary Report. *Journal of Urology*, **79,** 742.

ROSS, J. Cosbie, DAMANSKI, M., and GIBBON, N. (1960): Ureteric Reflux in the Paraplegic. *The British Journal of Surgery*, **47,** 636.

ROSS, J. Cosbie, GIBBON, N. O. K., and DAMANSKI, M. (1957): Recent Developments in the Treatment of the Paraplegic Bladder. *Lancet*, **ii,** 520.

ROSS, J. Cosbie, GIBBON, N. O. K., and DAMANSKI, M. (1958): Division of the External Urethral Sphincter in the Treatment of the Paraplegic Bladder. *British Journal of Urology*, **30,** 204.

SOULE, A. B., Jun. (1945): Neurogenic Ossifying Fibromyopathies. A Preliminary Report. *Journal of Neurosurgery*, **2,** 485.

OSTEOGENIC SARCOMA

An Analysis of Survival and its Relationship to Histological Grading and Structure

C. H. G. PRICE, BRISTOL, ENGLAND

Research Fellow in Pathology, University of Bristol

This analysis is an up-to-date review of work which has continued for the past nine years with steadily mounting material. The method used for assessing the grade of a tumour has already been described elsewhere (Price 1952, 1956), but is briefly recapitulated below.

Apart from the bearing which histological grading may have upon the estimation of prognosis, some means of measuring histologically the degree of malignancy of a tumour seems to be desirable because of the indecision still encountered in determining the relative merits of the two main forms of treatment of osteogenic sarcoma—radiotherapy and surgery. Although I am aware of the limitations of my technique for grading tumours, I believe that it does assist the clinician to decide the gravity of each sarcoma, to rationalise treatment, and to compile records that are more useful for comparison than those collected before its introduction. For the histologist, practice and familiarity with the system leads to added confidence in diagnosis, and in the author's hands the knowledge of the range of mitotic activity observed in osteogenic sarcoma has provided a useful and reliable criterion in the differential diagnosis of this tumour from reactive bone lesions.

The tumours in the present series include three variants which are classified in the records of the Bristol Bone Tumour Register under the generic term " osteogenic sarcoma," namely osteoblastic, chondroblastic and fibroblastic, this being in agreement with the usage of Coventry and Dahlin (1957). It is from the chondroblastic and fibroblastic types that most difficulties in designation arise.

The chondroblastic osteogenic sarcoma may contain much neoplastic cartilage showing calcification or ossification. It differs histologically from chondrosarcoma in showing much greater proliferation of tumour cells between areas of matrix. These cells may be atypical and undifferentiated, or may simulate osteoblasts or fibroblasts with greater or lesser formation of tumour osteoid or other intercellular material. This tumour is less uniformly cartilaginous than chondrosarcoma, in which proliferation of tumour cells in the matrix is relatively more characteristic. The fibroblastic osteogenic sarcoma has predominantly a fibrosarcomatous morphology, with much variation in the degree of collagenisation, some parts being densely sclerotic, others cellular and showing only a delicate network of fibrils. This fibroblastic sub-type appears relatively frequently as a complication of Paget's disease. Here and there however in these growths one finds cells resembling osteoblasts (or even chondroblasts) associated with irregular strands of osteoid material, occasionally with cartilage. From a study of sections of complete tumours I have come to regard primary pure fibrosarcoma in bone as a comparatively rare tumour, especially in older persons.

Although Luck (1957) and others have questioned the validity of regarding the presence of scattered islets of osteoid as a means of differentiating osteogenic sarcoma variants from pure fibrosarcoma of bone, there is usually no great difficulty in deciding whether such osteoid is proper to the tumour, or reactive in origin. Its form, location and relation to adjacent tumour cells moreover will often indicate if it is residual from invaded bone or reactive callus. The more common problem is to decide the nature of certain pleomorphic tumours which even in a large sample may be almost entirely devoid of matrix of any form. In the author's experience, these have usually been shown eventually to be osteolytic tumours composed o

very atypical osteoblastic cells which may show evidence of unusually rapid growth. Nevertheless, such pleomorphic neoplasms, usually essentially osteolytic, may occasionally show such a varied and atypical structure as to defy further classification beyond the broad term " osteogenic sarcoma."

MATERIAL AND METHODS

The present series of sarcomata is derived from the records of the Bristol Bone Tumour Register, and comprises the following: osteoblastic osteogenic sarcoma—forty-one tumours (47 per cent); chondroblastic osteogenic sarcoma—sixteen tumours (18 per cent); fibroblastic osteogenic sarcoma—thirty-one tumours (35 per cent) (Table I).

For grading, well preserved portions of tumour were selected from the specimens obtained at biopsy or excision. These were fixed, embedded in paraffin, sectioned and stained by conventional methods. Haematoxylin and eosin staining was commonly employed, often with a yellow substage filter used to increase the colour contrast of the cell nuclei. Mitoses in tumour cells were counted at a magnification of about 600, using a ruled screen in one

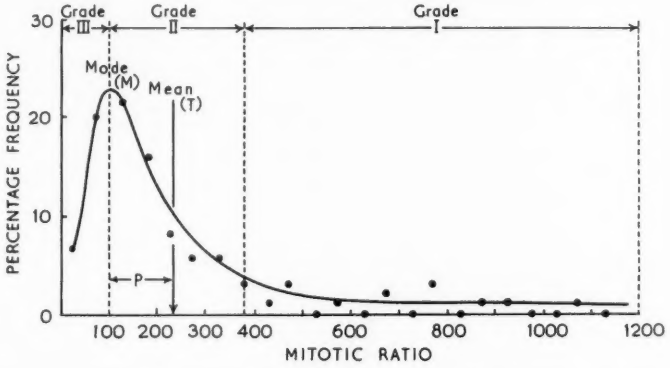


FIG. 1

The frequency distribution of the mitotic ratios of eighty-eight osteogenic sarcomata. The broken vertical lines indicate the subdivision by grades: Grade I (low malignancy): mitotic ratio *greater* than M+2P, where P is the interval between the modal and mean values for mitotic ratio. Grade II (average malignancy): mitotic ratio lies within the range of T±P. Grade III (high malignancy): mitotic ratio *less* than modal value.

eyepiece of a binocular microscope. At least 5,000 tumour cell nuclei were examined in each tumour, except in cases where mitoses were scanty, when two or three times this number of nuclei were examined. The mitotic ratio was calculated as the number of resting cell nuclei to one mitosis.

The three basic grades were defined as follows (Fig. 1): *Grade* 1—Low grade malignancy. This group includes all tumours in this series in which the mitotic ratio *exceeds* M+2P, where M is the modal value for the mitotic ratio and P is the interval between the mode and mean mitotic ratio. *Grade II*—Medium (or average) malignancy. The group consists of all tumours in which the mitotic ratio lies in the range of the mean value T of the mitotic ratio ±P. *Grade III*—High malignancy. This group includes all tumours in which the mitotic ratio is *less* than the mode M.

In an investigation of this kind it is essential to observe the following precautions: 1) All available sections of the tumour under examination should be studied, and the most ominous cellular areas selected for counting mitoses. 2) Areas selected for this purpose should be of good quality showing neither degeneration, necrosis, haemorrhage, sepsis nor mechanical damage. Tissue which has recently been irradiated should be avoided, as also should the

TABLE I

DETAILS OF THE BRISTOL SERIES OF OSTEOGENIC SARCOMATA

Number in register	Sex	Age at onset of symptoms (*years*)	Site	Type of tumour	Stage of tumour	Mitotic ratio	Grade	Treatment	Result	Period of survival after onset of symptoms (*months*)	Remarks
6	M	18	Femur	Osteoblastic	II	450	I	Deep x-ray. Amputation	Well	209+	
26	M	13	Humerus	Osteoblastic	II	677	I	Deep x-ray	Well	167+	
35	F	14	Femur	Osteoblastic	II	773	I	Deep x-ray. Amputation	Well	174+	
39	F	19	Femur	Osteoblastic	I	888	I	Amputation	Died	96	Diaphysial tumour
43	M	21	Femur	Osteoblastic	I	421	I	Amputation	Well	190+	
203	F	10	Humerus	Osteoblastic	I	727	I	Deep x-ray. Resection	Well	129+	Diaphysial tumour
317	M	89	Radius	Fibroblastic	I	926	I	Deep x-ray	Died	40	Paget's disease
595	F	21	Rib	Osteoblastic	I	1,100	I	Resection. Deep x-ray	Alive	71+	Has pulmonary metastases
664	M	8	Femur	Fibroblastic	II	492	I	Deep x-ray	Well	66+	
770	M	31	Navicular	Osteoblastic	II	750	I	Amputation	Well	56+	Lung resected for metastases
806	M	61	Tibia	Fibroblastic	II	694	I	Radio-cobalt	Died	51	Paget's disease
861	M	47	Rib	Osteoblastic	I	773	I	Resection. Radio-cobalt	Well	44+	
10	M	8	Humerus	Osteoblastic	II	190	II	Deep x-ray	Died	8	
61	M	15	Tibia	Osteoblastic	II	206	II	Deep x-ray. Excision	Died	20	
67	M	67	Femur	Fibroblastic	III	122	II	Amputation	Died	11	Post-irradiation sarcoma
70	M	58	Ilium	Osteoblastic	II	180	II	Deep x-ray to leg	Died	7	Paget's disease
81	M	22	Humerus	Osteoblastic	III	195	II	Deep x-ray	Died	27	
96	F	38	Femur	Fibroblastic	II	232	II	Deep x-ray to leg and chest	Died	48	Amputation delayed two years
102	F	70	Femur	Osteoblastic	II	132	II	Deep x-ray	Died	1[illegible]	
136	F	24	Femur	Chondro-blastic	I	113	II	Amputation. Deep x-ray to chest	Died	77	
143	M	9	Vertebra	Osteoblastic	II	153	II	Deep x-ray	Died	15	
145	M	26	Femur	Osteoblastic	I	108	II	Amputation	Died	15	Deep x-ray to chest and arm
169	F	62	Ilium	Osteoblastic	II	198	II	Deep x-ray	Died	12	
191	M	38	Femur	Osteoblastic	I	125	II	Excision. Amputation	Well	119+	
198	M	73	Femur	Osteoblastic	III	174	II	Amputation	Died	6	Paget's disease
213	M	16	Femur	Osteoblastic	II	125	II	Amputation	Died	30	
221	M	14	Femur	Osteoblastic	II	135	II	Deep x-ray. Amputation	Died	6	

Details of the Bristol series of osteogenic sarcomata stages, determined at the time of starting treatment are: I—confined to original bone; II—with local invasion; III—with regional metastasis; IV—with systemic metastasis

TABLE I—*continued*

DETAILS OF THE BRISTOL SERIES OF OSTEOGENIC SARCOMATA

Number in register	Sex	Age at onset of symptoms (*years*)	Site	Type of tumour	Stage of tumour	Mitotic ratio	Grade	Treatment	Result	Period of survival after onset of symptoms (*months*)	Remarks
226	F	64	Vertebra	Osteoblastic	I	168	II	None	Died	3	
299	M	67	Humerus	Fibroblastic	II	180	II	Amputation	Died	40	Paget's disease
337	F	36	Femur	Fibroblastic	II	134	II	Amputation	Well	115+	
338	M	60	Humerus	Fibroblastic	II	183	II	Deep x-ray	Well	106+	Paget's disease. Amputation delayed 5 years
406	M	66	Femur	Osteoblastic	II	250	II	Amputation	Died	41	Paget's disease
414	M	31	Humerus	Chondro-blastic	II	199	II	Deep x-ray. Resection. Amputation	Died	30	Deep x-ray to chest
416	M	69	Scapula	Fibroblastic	II	142	II	Deep x-ray. Resection	Died	18	Paget's disease
428	F	82	Femur	Osteoblastic	III	122	II	Amputation	Died	29	
450	M	60	Femur	Fibroblastic	II	140	II	Amputation	Died	29	
465	F	49	Femur	Fibroblastic	II	205	II	Amputation	Died	46	Deep x-ray to chest and skull
504	M	51	Femur	Fibroblastic	II	186	II	Resection. Amputation	Died	30	Paget's disease
512	F	76	Femur	Fibroblastic	II	323	II	Amputation	Well	81+	Paget's disease
522	F	45	Femur	Fibroblastic	II	104	II	Deep x-ray. Amputation	Died	21	
529	F	59	Femur	Fibroblastic	III	120	II	Deep x-ray to hip and thigh	Died	11	Paget's disease
538	M	72	Ilium	Chondro-blastic	II	310	II	Palliative	Died	11	Paget's disease
572	F	80	Femur	Fibroblastic	II	241	II	Palliative	Died	13	Paget's disease
579	M	20	Humerus	Fibroblastic	II	360	II	Deep x-ray. Amputation	Died	10	
586	F	53	Tibia	Fibroblastic	II	356	II	Amputation	Well	72+	
592	M	49	Femur	Osteoblastic	II	137	II	Deep x-ray to hip and thigh	Died	9	
691	F	18	Femur	Chondro-blastic	I	311	II	Amputation	Died	15	
705	F	57	Femur	Fibroblastic	II	130	II	Deep x-ray to thigh	Died	14	Paget's disease
737	F	9	Femur	Chondro-blastic	II	304	II	Amputation	Died	12	
741	F	67	Tibia	Fibroblastic	II	305	II	Amputation	Died	62	Deep x-ray to neck metastases
752	F	10	Tibia	Osteoblastic	I	288	II	Amputation	Died	16	
754	F	13	Humerus	Osteoblastic	II	165	II	Deep x-ray	Died	14	Deep x-ray to chest
765	M	73	Femur	Osteoblastic	II	185	II	Amputation	Died	7	Paget's disease

Details of the Bristol series of osteogenic sarcomata stages, determined at the time of starting treatment are: I—confined to original bone; II—with local invasion; III—with regional metastasis; IV—with systemic metastasis

TABLE I—*continued*

DETAILS OF THE BRISTOL SERIES OF OSTEOGENIC SARCOMATA

Number in register	Sex	Age at onset of symptoms (years)	Site	Type of tumour	Stage of tumour	Mitotic ratio	Grade	Treatment	Result	Period of survival after onset of symptoms (months)	Remarks
772	F	64	Femur	Fibroblastic	II	240	II	Deep x-ray. Amputation	Died	21	
803	F	85	Femur	Osteoblastic	II	290	II	Palliative	Died	3	Treated for fracture
847	F	36	Tibia	Fibroblastic	II	116	II	Amputation	Well	50+	
874	M	11	Humerus	Chondro-blastic	II	115	II	Deep x-ray. Amputation	Died	9	
883	M	41	Femur	Chondro-blastic	II	137	II	Palliative	Died	10	Paget's disease
894	M	19	Tibia	Chondro-blastic	II	127	II	Amputation. Deep x-ray	Died	16	Deep x-ray to groin, pelvis and shoulder
902	M	73	Femur	Fibroblastic	IV	280	II	None	Died	7	Paget's disease. Cardiac failure
910	F	42	Fibula	Osteoblastic	II	225	II	Deep x-ray	Well	67+	Amputation delayed 2 years
931	M	52	Humerus	Chondro-blastic	II	154	II	Radio-cobalt. Amputation	Died	25	Paget's disease
969	F	15	Femur	Chondro-blastic	II	198	II	Deep x-ray	Died	18	Nitrogen mustard
998	M	72	Femur	Chondro-blastic	II	209	II	Radio-cobalt	Died	20	
66	M	19	Tibia	Osteoblastic	IV	43	III	Deep x-ray to leg and chest	Died	11	
83	M	73	Mandible	Osteoblastic	II	65	III	Deep x-ray to mandible	Died	9	
94	F	73	Tibia	Osteoblastic	II	48	III	Deep x-ray	Died	5	Paget's disease
106	M	27	Maxilla	Osteoblastic	II	92	III	Palliative	Died	4	Fibrous dysplasia
155	M	71	Ilium	Osteoblastic	II	91	III	Deep x-ray	Died	11	Paget's disease
157	M	27	Femur	Fibroblastic	II	73	III	Deep x-ray	Died	59	Amputation delayed 1 year
163	M	13	Femur	Fibroblastic	II	88	III	Amputation	Died	5	
212	F	62	Calvarium	Osteoblastic	II	65	III	Deep x-ray	Died	7	Paget's disease
230	M	86	Femur	Fibroblastic	II	91	III	Palliative	Died	4	
250	M	14	Femur	Fibroblastic	II	83	III	Deep x-ray. Amputation	Died	18	
273	M	15	Humerus	Osteoblastic	II	46	III	Amputation	Died	15	
440	F	55	Femur	Fibroblastic	II	42	III	Amputation	Died	19	Paget's disease
516	M	75	Tibia	Osteoblastic	III	72	III	Deep x-ray. Amputation	Died	24	Paget's disease
588	M	59	Tibia	Chondro-blastic	I	83	III	Curettage. Amputation	Died	33	Recurrence in ilium

Details of the Bristol series of osteogenic sarcomata stages, determined at the time of starting treatment are: I—confined to original bone; II—with local invasion; III—with regional metastasis; IV—with systemic metastasis

TABLE I—*continued*

DETAILS OF THE BRISTOL SERIES OF OSTEOGENIC SARCOMATA

Number in register	Sex	Age at onset of symptoms (*years*)	Site	Type of tumour	Stage of tumour	Mitotic ratio	Grade	Treatment	Result	Period of survival after onset of symptoms (*months*)	Remarks
590	M	13	Femur	Fibroblastic	II	86	III	Deep x-ray	Died	13	
675	M	19	Humerus	Osteoblastic	II	61	III	Amputation	Died	11	Deep x-ray to chest
766	F	69	Tibia	Fibroblastic	II	54	III	Amputation	Died	7	Paget's disease
780	F	24	Femur	Fibroblastic	II	84	III	Amputation	Died	43	
830	M	16	Tibia	Chondro-blastic	II	71	III	Deep x-ray. Amputation	Died	14	Deep x-ray to metastases
918	M	16	Femur	Chondro-blastic	II	45	III	Deep x-ray. Amputation	Died	8	
958	M	16	Tibia	Chondro-blastic	II	57	III	Amputation	Died	15	
963	M	60	Femur	Osteoblastic	IV	69	III	Amputation	Died	5	Paget's disease
1,028	M	18	Femur	Chondro-blastic	II	59	III	Amputation	Died	7	
1,031	F	15	Rib	Osteoblastic	IV	69	III	Excision. Deep x-ray	Died	10	

Details of the Bristol series of osteogenic sarcomata stages, determined at the time of starting treatment are: I—confined to original bone; II—with local invasion; III—with regional metastasis; IV—with systemic metastasis

older parts of a tumour in which there is likely to be a high proportion of tumour matrix. 3) Should conflicting values for the mitotic ratio be obtained from several cellular areas of tumour tissue, the grade should be decided on the *minimum* figure so obtained on a full count of 5,000 cell nuclei.

With few exceptions, the grading of the present series of eighty-eight tumours was done while the patient was alive. No attention was paid to history, radiographic appearances or findings at operation, although this information was known at the time of grading. Hence, although the study cannot be regarded as having been performed "blind," the data given for the connection between grade and survival could not well have been cumulatively deduced from such details—particularly in respect of those tumours designated Grade II and Grade III. No attempt was made at any time to adjust the histological grade to conform better with the known details of the tumour or of the patient.

RESULTS

Subdivision into three basic grades of malignancy—The mitotic ratio for each tumour is shown in Table I. The mean ratio for the whole series was 231.

The frequency distribution of the mitotic ratios of these eighty-eight sarcomata is plotted in Figure 1. This curve should be compared with that showing the frequency distribution of survival times in months from the date of the presenting symptom (Fig. 2). These two curves are rather similar and have a lognormal distribution around the mode. They indicate the rationale of the subdivision of the present series of tumours into grades of low, medium and high malignancy. Further subdivision serves no useful purpose.

On reviewing this work it was found that the method of grouping by grades roughly corresponded with that which had formerly been adopted (Price 1952) and which had been

based upon consideration of the known survivals of the original thirty-seven sarcomata in the 1952 analysis.

The distribution of the tumours in this series according to grade is shown in Table II.

Correlation between mitotic ratio and survival—When this method of grading osteogenic sarcomata was first devised, the function chosen to indicate mitotic activity was selected in

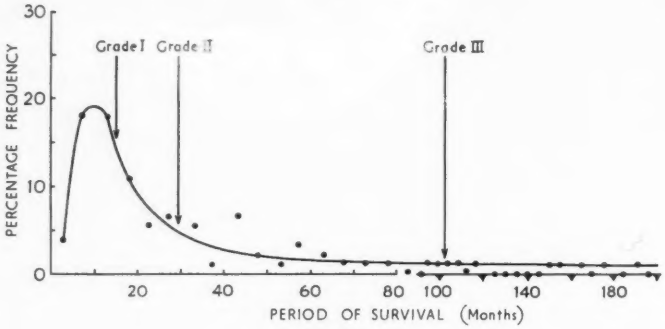


FIG. 2

The frequency distribution of survivals of eighty-eight patients with osteogenic sarcoma. The vertical arrows indicate the mean survival in the three grades. Note break in the time scale.

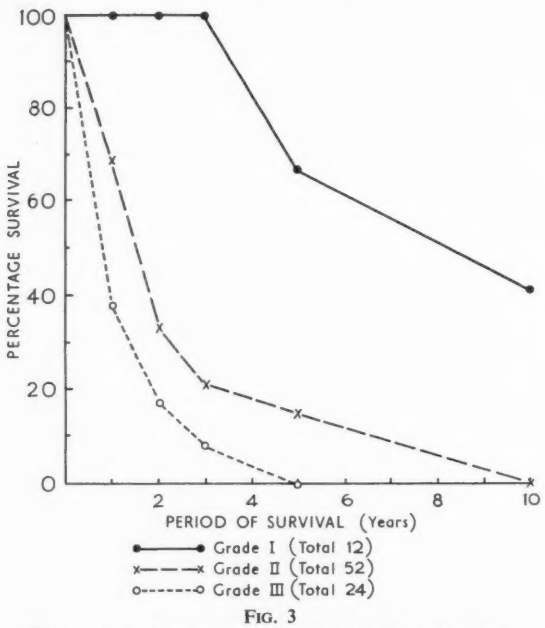


FIG. 3

The survival of patients according to the grade of tumour.

order that it might possibly give a positive correlation with survival rather than a negative The correlation coefficient " r " has been calculated on several occasions for the two variate mitotic ratio and months survived by each patient. The results are shown in Table III.

Furthermore, the mean mitotic ratio of twenty-eight tumours in patients who survive for longer than the average of thirty-six months for the whole series was 413, as compare

with a mean mitotic ratio of 156 for sixty tumours in patients who survived for less than this period. The difference of the mean mitotic ratio for these two classes is statistically significant (p being less than 0·001).

The correlation coefficients for the same two variates have also been calculated separately for the three grades of tumours (Table IV).

Only in Grade III does any positive relationship still exist between the mitotic ratio and survival. There are a number of reasons which may account for this: 1) Treatment has obviously

TABLE II

DISTRIBUTION OF TUMOURS ACCORDING TO GRADE

Grade	Number of tumours	Percentage	Mitotic ratio
I	12	14	Over 370 (over M+2P)
II	52	59	100 to 370 (T±P)
III	24	27	Under 100 (less than M)

TABLE III

CORRELATION BETWEEN MITOTIC RATIO AND SURVIVAL

Year	Total number of tumours	r	p
1952	37	+0·62	Less than 0·01
1956	66	+0·53	Less than 0·01
1959	88	+0·59	Less than 0·01

TABLE IV

CORRELATION BETWEEN MITOTIC RATIO AND SURVIVAL IN THE THREE GRADES

Grade	Number of tumours	r	p
I	12	−0·54	Less than 0·1
II	52	+0·04	Greater than 0·1
III	24	+0·18	Greater than 0·1

had the greatest effect among the small group of Grade I cases, five of whom appear early in the series and have hence been under observation for long periods. 2) Factors other than histological grade, for instance, site, stage and size of the primary tumour, and complications such as fracture or sepsis, may be influencing the outcome. Some indication of this effect may be shown by excluding such biased cases, when recalculation of " r " gives the increased value of +0·32 for Grade III. 3) Thirty-two per cent of the patients included in this series have been under observation for less than five years (Case 705 onwards). 4) Within the reduced range of the mitotic ratio for Grades II and III considered separately, the personal and sampling errors in evaluating the mitotic ratio assume a much greater relative significance.

TABLE V

RELATION BETWEEN HISTOLOGICAL GRADE AND SURVIVAL

Grade	Number of tumours	Mean mitotic ratio	Survival (*months*)				Medians (*in months*)	
			Mean	s	Difference of means	s_D	Survival	Mortality
I	12	697	108	62·7	79	18·53 (p less than 0·001)	78	40
II	52	194	29	28·6				
II	—	—	29	28·6	14	4·81 (p less than 0·01)	17	15
III	24	68	15	13·3				
All	88	231	36	—	—	—	17	14

This Table compares the mean survival of patients with Grade I tumours with that of patients with Grade II tumours; likewise for patients with Grade II and Grade III tumours. These observed differences of means are shown in column 6. In column 7 is shown the statistical significance based upon calculated values of "t." $\left(\frac{\text{Difference of means}}{\text{Standard deviation of difference of means } (s_D)}\right)$.

TABLE VI

RELATION BETWEEN HISTOLOGICAL GRADE AND PERCENTAGE SURVIVAL

Grade	Percentage survival					Survival per cent of expectation of life (actuarial)
	1 year	2 years	3 years	4 years	5 years	
I	100	100	100	67	41	17·7
II	69	33	21	15	nil	7·5
III	38	17	8	nil	nil	3·6
All	57	33	25	15	5	8·5

TABLE VII

RELATION BETWEEN SURVIVAL AND TYPE OF TUMOUR (LONG BONES ONLY)

Type of tumour	Number of tumours	Percentage	Mean survival (*months*)	s	Percentage survival		
					3 years	5 years	10 years
Osteoblastic	29	40	50	62·8	31	28	21
Fibroblastic	28	40	39	29·9	50	18	0
Chondroblastic	14	20	20	18·2	7	7	0
All types	71	—	39	—	34	20	8

The relationship between histological grade and survival—The relationship between grade and survival is shown in Tables V and VI.

The differences of the mean survivals between Grades I and II, and between Grades II and III, are statistically significant; moreover, it is only in Grade I that there is any marked difference between median survival and median mortality.

The survival data are also plotted in Figure 3.

Negative correlation between mitotic ratio and survival—There were eighteen patients in whom this correlation was negative. Nine survived longer than expected; nine did not survive as long as expected.

Patients surviving longer than expected—These were Cases 136, 157, 191, 337, 338, 465, 780, 847 and 910. The tumour in two of these patients was osteoblastic, in one chondroblastic, and in six fibroblastic in type (Table I). Even with this very small group certain trends may be discerned: 1) Most of the tumours were the fibroblastic variant. 2) All nine tumours were situated in long bones and were treated by amputation (in one instance, Case 338, delayed for five years after radical radiotherapy). 3) It is unusual for a patient with a Grade III tumour to survive longer than the mean for the whole series of thirty-six months. 4) None of these patients were adolescents, and seven were middle-aged. At this period of life there is relative metabolic stability and maturity of connective tissue; possibly the prognosis in this age group is somewhat better in each grade than in either youth or old age. 5) Of the whole series of eighty-eight patients, eighteen (22 per cent) were between twenty-five and fifty-four years old;

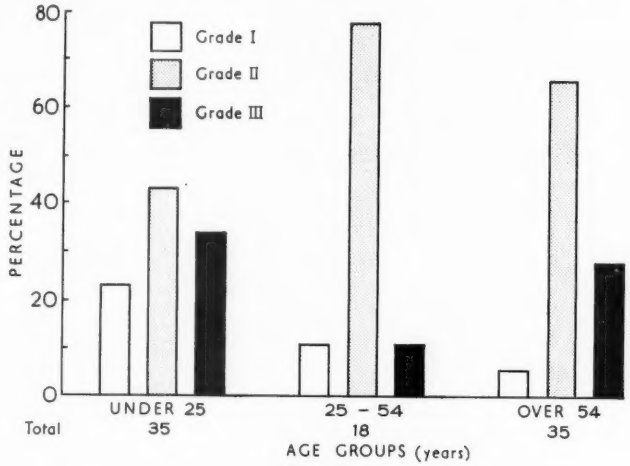


FIG. 4

The relation between age and grade in the Bristol series.

sixteen of these had tumours of long bones. There is thus a good chance of a person in this age group surviving longer than the mean thirty-six months irrespective of the grade of tumour. 6) Grade III tumours seem to be relatively uncommon (11 per cent) in middle life (Fig. 4).

Patients not surviving as long as expected—These were cases 538, 572, 579, 691, 737, 752, 772, 803 and 902. In four of these nine patients the tumours were fibroblastic, in three chondroblastic and in two osteoblastic (Table I). The following points are perhaps noteworthy: 1) The nine patients were all either juveniles or aged. 2) The nine tumours were all of Grade II, eight being situated in long bones. Only five of these were treated by amputation, three receiving only palliative treatment. 3) Two tumours were complicated by Paget's disease, and one (Case 579) by a severe fracture and sepsis. 4) Of the three juveniles (Cases 691, 737, 752), two had chondroblastic tumours, which in this series at least seem to have a graver prognosis than the two other types of osteogenic sarcoma.

The relationship between survival and type of tumour—The analysis of the connection between these two factors in tumours *of long bones only* is shown in Table VII. The differences in the mean survival times are not statistically significant except for the comparison between the fibroblastic and chondroblastic tumour groups, where p is less than 0·001. The Table does serve however to indicate the tendency for short survival of patients with this latter type of tumour.

The relationship between survival and site of the tumour—The present series is too small to permit any useful analysis of prognosis related to the site of the growth, with the possible exception of the group of forty-five femoral tumours. The mean survival of these patients was forty months, and included nine five-year survivors (20 per cent). This figure seems to reflect the general run of experience elsewhere. No patient with a tumour of skull, axial or girdle skeleton has survived for more than eighteen months irrespective of grade, and the mean survival of ten such patients was only ten months.

DISCUSSION

The method of grading osteogenic sarcomata described here is primarily conceived as an attempt to measure the degree of malignancy of individual tumours. Whilst this quantitative method does not run entirely counter to the histo-morphological grading as described by Broders (1940) and applied to osteogenic sarcomata by Coventry and Dahlin (1957), it has been found that tumour structure is usually so varied that the morphological method is deceptive and unreliable. In this conclusion the author is in agreement with other workers. Furthermore, because these neoplasms—at least those of slower growth and lower grade—tend to have a characteristic micro-anatomy, any method of grading which does not take cognisance of this fact is likely to be fallacious.

The basic assumptions on which the present method relies are twofold: 1) That cell proliferation in osteogenic sarcomata is mainly mitotic; this does not obtain in all connective tissue growths, for example, chondrosarcoma. 2) That it is generally agreed that the presence of many mitotic figures in any neoplasm indicates high malignancy.

The present work then attempts to define the range of mitotic activity in osteogenic sarcoma, to subdivide this range by means of some intrinsic features, and to indicate the relationship of the three defined tumour grades to survival.

Owing to the differing usages of bone tumour nomenclature, it has been necessary to indicate what types of sarcoma are here included under the heading " osteogenic." The wide variation in structure of this sarcoma is well known—quite how wide is perhaps a matter for more personal opinions. Having decided the admissible range of histological structure of osteogenic sarcoma, further analysis by structural types seems to offer little, save to indicate the tendency for a poorer prognosis for the chondroblastic variant. The author has repeatedly observed the marked vascularity associated with cartilage resorption—for instance in the normal epiphysial growth plate. This factor may possibly be of importance in the rapidity and ease with which chondroblastic tumours metastasise.

The tumours of older persons show relatively less matrix formation than those typically seen in juveniles. For this reason, the radiographic appearances of osteogenic sarcoma in persons over sixty years are predominantly those of osteolysis and bone destruction. Not infrequently at this age the presenting symptom is pathological fracture, which is followed by the revelation of the underlying sarcoma. This sequence is distinctly uncommon in adolescents.

When the original small group of thirty-seven tumours was studied in 1952 some difficulty was encountered in deciding how to define the three basic grades of malignancy. At that time the only quantitative landmarks discernible were the limits of the range of the mitotic ratio, and the mean value for the thirty-seven tumours (250). Using this mean as the central point, Grade II was then defined as the mean mitotic ratio plus or minus a range of 150. It may now be noted that greater significance is attached to the modal value for the mitotic ratio, which is approximately 100. The further significance of the mode is that it marks the point at which malignancy of these tumours ceases to increase as a logarithmic function of their frequency, but instead becomes inversely proportional to that function. Moreover, clinically this modal point is a convenient mark for separation of Grades II and III—those sarcomata in which respectively some benefit may and may not be reasonably expected to follow treatment.

It is interesting to compare the results in the present series so graded with the data published by Coventry and Dahlin (1957) (Table VIII).

TABLE VIII

DISTRIBUTION OF GRADES AND FIVE-YEAR SURVIVAL
(Price 1961, Coventry and Dahlin 1957)

	Grade	Percentage of total	Percentage survival (5 years)	Total patients
Price 1961	I	14	67	88
	II	59	15	
	III	27	0	
Coventry and Dahlin (1957)	1 and 2	18·8	25·4	430
	3	54·4	17·6	
	4	26·7		

TABLE IX

RELATION OF MITOTIC RATIO TO AGE

Age (*years*)	Number of patients	Mean mitotic ratio
Under 25	35	267
25 to 54	18	228
Over 54	35	197

In the two series the proportions of tumours in each grade agree tolerably well if it be assumed that Grades 1 and 2 of the American series roughly correspond with the British Grade I. The survival rate of patients with tumours of Grades II and III combined (15 per cent) is similar to that of patients with tumours of Grades 3 and 4 combined. It seems likely that the Price Grade I is rather more restrictive than the Coventry and Dahlin Grades 1 and 2 combined but that it is a good deal wider than Grade 1 alone, because only 1·6 per cent of tumours were included in this grade. The British series is undoubtedly weighted in the lowest grade by the favourable course of five young patients early in the series who all had osteoblastic tumours of long bones. One other juvenile in this group who had an osteoblastic tumour of the femur succumbed to pulmonary metastases after eight years (Table I).

There is one obvious difference between the two series compared above: in southern England the incidence of osteogenic sarcoma is probably highest amongst older persons, not amongst adolescents (Price 1955, 1958). This is largely due to the frequency of Paget's sarcoma in this part of the country. It may however be added that there appears to be no significant difference in the average mitotic ratio of Paget's sarcomata as compared with that of uncomplicated osteogenic sarcomata in patients of the same age group.

Age and malignancy—In Figure 5 the relation of age to degree of malignancy for this series and for that of Coventry and Dahlin (1957) is shown. In my view the dividing line of age should be twenty-four not twenty, so as to include all tumours caused by an aberration of skeletal growth. There is a general broad similarity of results in the two series, but in the British series the degree of malignancy tends to increase rather than decrease as a function of age. This is also suggested by the decrease in the mean mitotic ratio with age (Table IX).

The addition to the British series of a further 14 per cent of middle-aged patients in order to make the age distribution more similar to that in the American series would raise still higher the proportion of tumours of Grades II and III, so accentuating this difference.

Apart from differences of grading technique, and the relatively small number of patients in the British series, it seems that there are racial differences in the etiology of osteogenic sarcoma. For example, Paget's sarcoma is common in England, less common in the United

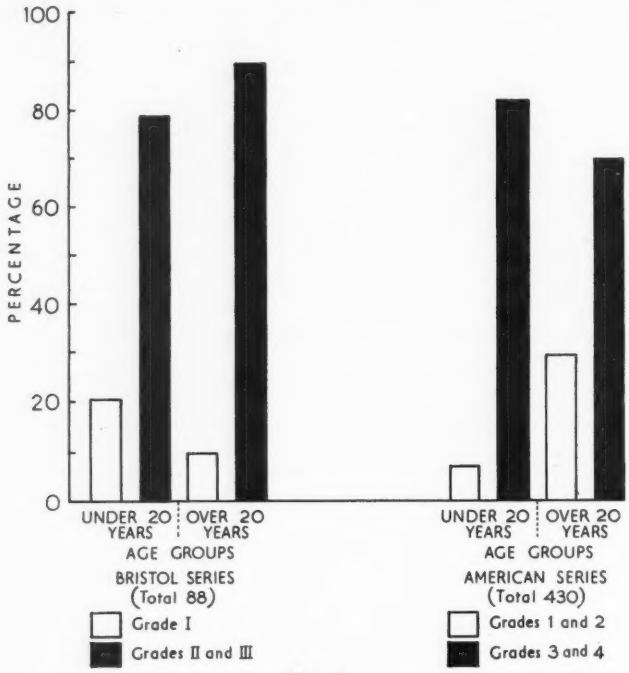


FIG. 5

The relation between age and tumour grade, in the British (*left hand*) and American (*right hand*) series. The data relating to the American series are drawn from Coventry and Dahlin (1957).

States (probably by a factor of ten or more), and rare in Scandinavia. Familial examples of Paget's disease are uncommon even in England, but are not so uncommon as familial cases of osteogenic sarcoma. Genetic studies of the blood groups have shown a surprising broad stability of certain European populations, hence it is interesting to speculate to what extent the mixing of racial strains may have contributed to the reduced incidence of Paget's sarcoma in the United States, or to what extent this might be due to heterosis—perhaps a " distinction without a difference."

SUMMARY

1. The relationship between histological grading and survival has been studied in a consecutive series of eighty-eight patients with osteogenic sarcomata. The grading has been based entirely upon the mitotic activity of these tumours.

2. There is a positive correlation (" r "$=+0{\cdot}59$) between the two variates mitotic ratio and survival in months.

3. The frequency distributions of the mitotic ratios and survivals are similar and, from the observed range, mean and mode of mitotic ratios a more precise definition is proposed for the terms " low," " medium " and " high " malignancy.

4. Comment is made on those patients (20 per cent) in whom actual survival was very different from that expected on the basis of histological grading.
5. The five-year survival rates were: Grade I—67 per cent, Grade II—15 per cent, Grade III—nil, all—17 per cent. For the whole series the average survival period from the time of the presenting symptom was thirty-six months.
6. The five-year survival rate for forty-five tumours of the femur was 20 per cent; no further analysis by sites is attempted.

The author wishes to express his thanks for the contributory assistance of fellow members of the Panel of the Bristol Bone Tumour Registry, who by referring their cases for discussion have supplied the major part of the present series of sarcomata, thus providing the foundation of the present study. Added to these local patients are a certain number which have been derived from the following centres: Bedford, Birmingham, Coventry, Gloucester, Exeter, Northampton, Plymouth, Salisbury, Taunton, Truro, Utrecht (Holland) and York. To the surgeons and pathologists of these several cities who have thus helped to provide material the author is much indebted, as also to Mr G. M. Clarke for his advice with the statistical treatment of the analyses.

The Bristol Bone Tumour Registry together with the present investigation are supported by generous grants from the British Empire Cancer Campaign, to whom the author's thanks for a personal grant are also due.

REFERENCES

BRODERS, A. C. (1940): The Microscopic Grading of Cancer. In *Treatment of Cancer and Allied Diseases*. Edited by G. T. Pack and E. M. Livingston. Volume 1, pp. 19–41. New York: Paul B. Hoeber, Inc.

COVENTRY, M. B., and Dahlin, D. C. (1957): Osteogenic Sarcoma. *Journal of Bone and Joint Surgery*, **39–A,** 741.

LUCK, J. V. (1957): *In* Discussion on Paper by Coventry and Dahlin. *Journal of Bone and Joint Surgery*, **39–A,** 757.

PRICE, C. H. G. (1952): The Grading of Osteogenic Sarcoma. *British Journal of Cancer*, **6,** 46.

PRICE, C. H. G. (1955): Osteogenic Sarcoma: an Analysis of the Age and Sex Incidence. *British Journal of Cancer*, **9,** 558.

PRICE, C. H. G. (1956): The Grading of Osteogenic Sarcoma, and its Bearing upon Survival and Prognosis. *Journal of the Faculty of Radiologists*, **7,** 237.

PRICE, C. H. G. (1958): Primary Bone-forming Tumours and their Relationship to Skeletal Growth. *Journal of Bone and Joint Surgery*, **40–B,** 574.

A CASE OF NEUROPATHIC ARTHRITIS CAUSED BY INDIFFERENCE TO PAIN

H. VAN DER HOUWEN, LEIDEN, HOLLAND

From the Anna-Kliniek voor Orthopaedie, Leiden

Severe degenerative joint changes can be caused by lack of the protection afforded by the sense of pain. Usually the loss of pain sense is known before the joints are damaged. Recently we have investigated a man with a generalised analgesia and neuropathic arthritis of several joints, who had only a localised sensory disturbance at the age of thirteen when the first joint became affected.

CASE HISTORY

A boy, born in 1935, was seen at another hospital in 1948 because of a swollen right knee. The first radiographs showed no bony changes (Figs. 1 and 4), but after a further six months he still had a hydrarthrosis and the lateral femoral condyle was seen to be roughened (Fig. 2). The diagnosis remained obscure and an arthrotomy revealed degenerative arthritis only. A few months later, suffering from difficulty in walking because of instability of the knee, he was admitted to the neurological department of a teaching hospital; here he was observed for over three months. The senses of pain and temperature were found to be abnormal only in the right leg below the knee, although it was noted that the interpretation was difficult. Lumbar puncture and serological tests for syphilis gave normal results. On discharge he was regarded as having an arthropathia of unknown origin, and was advised to wear a protective brace, but this he refused.

In 1953 he suffered a blow on his left forearm, but continued to work. The same evening the arm was swollen and he was found to have a fracture of the ulna. Although the arm was immobilised in a plaster for six months a pseudarthrosis developed; at first the elbow joint was normal but gradually destruction occurred (Figs. 6 and 7). The arm was never painful.

In 1959 he jumped from a height of two feet and his left knee gave way, having sustained a painless compression fracture of the medial femoral condyle (Fig. 5). He was ordered complete rest but he ignored this advice, so he was admitted to this hospital. Aspiration of the knee was performed repeatedly, and up to 400 millilitres of sero-sanguinous fluid were removed each time. The right knee was now found to be completely destroyed, with the distal end of the femur lying on the medial side of the tibial condyle, and five centimetres of shortening of the leg. Considerable calcification round the joint was visible in the radiographs (Fig. 3) but, surprisingly, the knee was stable.

The lumbar spine showed scoliosis to the right, with narrowing of the lumbo-sacral disc and secondary bony changes (Fig. 8), but he had no complaints about his back. The destruction of the left elbow had increased; the radial head was dislocated and considerably displaced. The pseudarthrosis in the ulna was still present, and the bone was worn down at the fracture site (Fig. 7).

Employed as a wood sawyer, his intelligence was somewhat below the average, although he was alert. He was cooperative in the hospital, but from his history it seems that he was rather indifferent to medical advice and failed to keep his appointments. He could not remember having had headaches or abdominal pains, but he had suffered from toothache. He was sensitive to tickling and itching. He said that he could easily walk barefoot on gravel and that he could touch nettles without pain. In his youth he had suffered from inflamed toes but now there were no signs of trophic disturbances. He bore scars over the medial malleoli where he had kicked himself.

FIG. 1

FIG. 2

Figure 1—Right knee in 1948. Figure 2—Right knee in 1949.

FIG. 3

Right knee in 1959.

FIG. 4

FIG. 5

Figure 4—Left knee in 1948. Figure 5—Left knee in 1959.

FIG. 6 FIG. 7
Figure 6—Left arm in 1956. Figure 7—Left arm in 1959.

Family history—The patient was the youngest of a family of nine children of whom five had died in childhood after illnesses of short duration. One sister who died in 1940 at the age of seven probably also suffered from indifference to pain as she sometimes jumped from a table and landed on her knees without signs of discomfort. She also used to scratch at little cuts which were apparently painless.

Neurological investigations—The indifference to painful stimuli was generalised, including the oral mucosa. He felt pinpricks as such, but said it was not painful. He could discriminate between sharp and blunt, but would make some mistakes when the point of a pin was applied lightly or when the head was applied firmly. Pulling on his hairs, muscle-ischaemia—five minutes at a pressure of 180 millimetres of mercury—and severe electrical stimulation were endured with a smile. Pinching the calcaneal tendon and the testes (as well as aspirating the knee with a thick needle) were not painful, and produced no dilation of the pupils. A ridge in the plaster cast immobilising his left knee caused an excoriation which was hardly noticed by the patient.

FIG. 8
Lumbar spine in 1959.

The pupils showed a tonic contraction to light, corneal sensation was diminished and the reflex was sluggish; touching the right cornea was less unpleasant than the left. Vision, the visual fields, hearing and smell were normal. There was no wasting or fasciculation of muscles, in which tone and power were normal, the abdominal reflexes were present and equal, and the tendon reflexes were normal except in the right knee which may have been because of the deformity of the joint; no other abnormal reflexes were found. Light touch, two-point discrimination, vibration and postural sense and stereognosis were normal. The patient correctly located the point of stimulation and there was no confusion between right and left. Temperature sense was undisturbed, with discrimination between slight differences in temperature (he had never suffered painless burns and he was careful with heat). The electro-encephalograph was normal.

Other investigations—The alkaline phosphatase was slightly raised (18,2 King-Armstrong units); the blood-Kolmer and V.D.R.L.* reactions were negative.

* The standard cardiolipin test of the United States Venereal Diseases Reference Laboratory.

DISCUSSION

The severe joint changes in this patient were occasioned by an isolated loss of pain sensation. Trophic disturbances are not a part of this condition and are apparently not a requisite for the development of neuropathic joints. According to Silverman and Gilden (1959) forty-six such cases have been published; when this analgesia is discovered in childhood it is called congenital indifference to pain. It is remarkable that in this patient the sensory defect was found so late. Even on thinking back his parents can remember only a few occasions on which he may have shown more than usual insensitivity to pain.

The lesion is thought to be somewhere in the thalamic region of the brain, but there have been no reports on the pathology of this condition. In hereditary sensory radicular neuropathy, in which there are also defects of other senses, lesions have been found in the dorsal root ganglia with atrophy of the ascending tracts.

The differential diagnosis of a neuropathic joint includes tabes dorsalis, syringomyelia and diabetic neuritis. Analgesic states can be found in mental retardation, hysteria and schizophrenia. Schilder and Stengel (1931) described asymbolia for pain caused by an acquired lesion in the posterior parietal region of the dominant hemisphere. A similar state can be caused by double division of the spino-thalamic tract and by frontal leucotomy. The other affections of bone seen in patients with indifference to pain are osteomyelitis and deformities due to malunited fractures.

SUMMARY

1. A twenty-five-year-old man who developed neuropathic arthritis because of indifference to pain is described. He presented at the age of thirteen with degenerative changes in the right knee with analgesia only in the right leg below the knee.
2. A painless fracture of the ulna developed non-union and was associated with a neuropathic arthritis of the elbow joint.
3. Later the other knee and the lumbo-sacral spine became affected. The analgesia became generalised with all other senses staying intact. Apart from tonic pupillary reactions and diminished corneal sensibility no other neurological signs could be found.
4. A deceased sister may also have suffered from the same condition.

I am very grateful to Dr J. D. Mulder, Dr F. Duyfjes, and to Dr F. Steenwinkel, of the Anna-Kliniek, for their helpful criticism and advice.

REFERENCES

CAMBIER, J., and LEFEBVRE, J. P. (1959): L'indifférence congénitale à la douleur. *Presse Médicale*, **67,** 1,551.
CRITCHLEY, M. (1956): Congenital Indifference to Pain. *Annals of Internal Medicine*, **45,** 737.
FANCONI, G., and FERRAZZINI, F. (1957): Kongenitale Analgie. *Helvetica Paediatrica Acta*, **12,** 79.
JÉQUIER, M., and DELLER, M. (1956): L'indifférence congénitale à la douleur. *Confinia Neurologica*, **16,** 207.
JEWESBURY, E. C. O. (1951): Insensitivity to Pain. *Brain*, **74,** 336.
MURRAY, R. O. (1957): Congenital Indifference to Pain with Special Reference to Skeletal Changes. *British Journal of Radiology*, **N.S. 30,** 2.
ORTIZ DE ZARATE, J. C. (1955): Analgésie généralisée congénitale. *Encéphale*, **44,** 414.
PETRIE, J. G. (1953): A Case of Progressive Joint Disorders Caused by Insensitivity to Pain. *Journal of Bone and Joint Surgery*, **35–B,** 399.
SCHILDER, P., and STENGEL, E. (1931): Asymbolia for Pain. *Archives of Neurology and Psychiatry*, **25,** 598.
SILVERMAN, F. N., and GILDEN, J. J. (1959): Congenital Insensitivity to Pain: a Neurologic Syndrome with Bizarre Skeletal Lesions. *Radiology*, **72,** 176.

PROGRESSIVE FIBROSIS OF THE VASTUS INTERMEDIUS MUSCLE IN CHILDREN*

A Cause of Limited Knee Flexion and Elevation of the Patella

O. HNĚVKOVSKÝ, PRAGUE, CZECHOSLOVAKIA

From the Second Clinic for Orthopaedic Surgery, University of Prague

In the nine years from 1951–59 we have observed twelve children—eight girls and four boys—in whom a marked limitation of flexion of the knee joint developed in a relatively short time. The children were between one and seven years old (Table I). The parents of all of them stated that at the beginning the knee movements were restricted only very little and that it was only in the course of time that they grew more and more limited. In no case was there a previous history of trauma or of inflammation of the joint or of neighbouring tissues; the knee was never swollen, warm or painful. In the youngest children the limitation of movement was usually noticed because the child could not sit on the chamber pot in the normal way; in older children insufficient flexion prevented them from squatting. When walking, the children did so with an extended knee even if movement of the knee was still partially preserved.

The initial diagnoses in most cases were those of trauma or of inflammation, although all the usual symptoms of these conditions were missing. In one case, probably because the patellar tendon reflex was absent, the condition was thought to be a post-poliomyelitic contracture. The children were initially treated by plaster splints or, more often, by manipulation and physiotherapy. In no case did such treatment effect improvement; in fact, mobility steadily decreased.

FIG. 1

FIG. 2

Figure 1—Case 4. Girl aged four. Note elevation of right patella. Figure 2—Case 3. Girl aged nine. Marked elevation of right patella.

All the children were first examined by us after having been treated in different ways. We also attempted once more to mobilise the knees by manipulation, splints, massage, electrotherapy and so on, but we did not succeed even temporarily in improving the flexion. On the contrary, we found that children who were temporarily discharged from our clinic and who returned some months later, always had even a more limited flexion than during their first hospitalisation (Cases 1, 3, 4, 9, 10, 11, 12).

CLINICAL PICTURE

In eight children the condition affected only one knee and in four children there was a limitation of flexion of both knees. In one girl (Case 3) so affected there was also partial rigidity of some muscles of the leg and foot like that seen in an incomplete syndrome of arthromyodysplasia (arthrogryposis). This case is therefore rather different from the others in its suggestion that there may be a connection between arthromyodysplasia and our cases of progressive fibrosis of the vastus intermedius. There is of course the difference that in

TABLE I
ANALYSIS OF MATERIAL

* Submitted for publication January 1960.

TABLE I

ANALYSIS OF MATERIAL

Case number	Sex	Side affected	Other congenital anomalies	Age at onset (*years*)	Knee flexion		Previous treatment	Age at operation (*years*)	Movement after operation		Investigations	Text figures
					Age (*years*)	Range (*degrees*)			Time (*years*)	Range (*degrees*)		
1	F	Left	—	1	3 6	90 5	Manipulation. Physiotherapy	6	3	80	Electromyography. Measurement of chronaxie	—
2	M	Right	—	1	2	15	Manipulation. Physiotherapy	2	3	130	Electrical excitability at operation	11
3	F	Right (left knee flexion 120 degrees)	—	1	6 9	90 35	Manipulation. Physiotherapy. Balneotherapy	9	2	90	Electromyography. Electrical excitability at operation	2, 9
4	F	Right (left knee flexion 135 degrees)	Congenital dislocation of hip	1¾	2 4 6	110 90 40	Manipulation. Plaster splintage. Physiotherapy	6	2	90	Electromyography	1
5	F	Left	—	2	6	10	Manipulation on three occasions. Plaster splintage	6	3	100	Electromyography. Measurement of chronaxie. Electrical excitability at operation	—
6	F	Left	Congenital coxa valga	2½	2½	40	As for specific arthritis	2½	1	105	Electromyography. Histology	—
7	M	Right	Congenital dislocation of hip	3	4	35	Plaster splintage. Physiotherapy	4	1	75	Electrical excitability at operation. Measurement of chronaxie. Histology	—
8	M	Left	—	3	9	15	Plaster splintage. Manipulation	—	—	—	Electromyography. Measurement of chronaxie. Histology	3
9	F	Left	—	3	3½ 5	90 10	Physiotherapy	5	2	95	Electromyography. Measurement of chronaxie	10
10	M	Left	—	3½	4 4½	65 45	As for specific arthritis	4½	1	90	Electromyography. Histology	—
11	F	Right (left knee flexion 125 degrees)	—	6	7 8	90 55	Manipulation. Plaster splintage. Physiotherapy	—	—	—	Electromyography	—
12	F	Right (left knee flexion 100 degrees)	—	7	4 13	100 10	As for poliomyelitis	13	1	90	Electromyography. Measurement of chronaxie	4

this patient the condition of the knee joint constantly deteriorated, which does not occur in arthromyodysplasia.

The appearance of the affected knee was not markedly different from that of the other one. In older children the patella was situated somewhat higher and more laterally than normal (Figs. 1 and 2). In the older children, where the pathological process had been going

FIG. 3

Figure 3—Case 8. Boy aged nine. Radiographs showing elevation of left patella.

FIG. 4

Figure 4—Case 12. Girl aged thirteen. Radiographs showing elevation of left patella.

on for some time, there was wasting of the thigh of about half an inch (1 to 1·5 centimetres). There was no wasting of the calf, except in the case of the girl (Case 3) with obvious affection of the muscles of the lower limb.

On the front of the thigh there could be felt a tendon-like structure attached to the patella. This was most easily felt when the knee was flexed as far as possible. There were no changes in the lateral parts of the extensor apparatus of the knee.

The patellar reflexes were diminished or even absent, and the knee flexor reflexes were weaker than normal. All the other reflexes of the lower extremity and reflexes elsewhere in the body were normal.

The knee movements were limited in a characteristic way. During an active or passive attempt at flexion the movement was always stopped exactly at one point, and quite suddenly, as if a solid obstacle were in the way. Movement was not limited by pain and even forcible attempts at flexion were not painful. It seemed that the block was caused by the middle part of the tendon of the quadriceps muscle. As extension of the hip joint was not limited, it appeared that flexion of the knee was limited by some condition affecting the vastus intermedius rather than the rectus femoris.

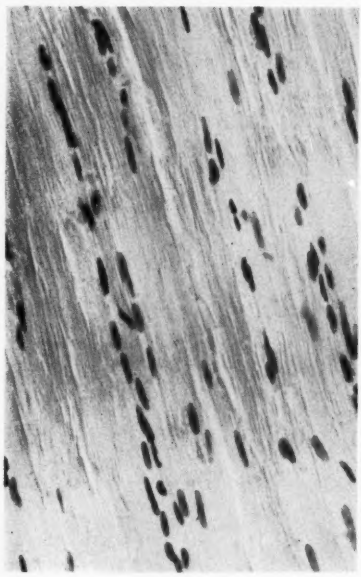


FIG. 5

FIG. 6

Histological appearances. Figure 5—Increase in number of endomysial and sarcolemmal nuclei. (Haematoxylin and eosin, ×270.) Figure 6—Proliferation of endomysial and sarcolemmal nuclei with invasion of sarcoplasm. Degeneration of muscle fibres with partial loss of striation. (Haematoxylin and eosin, ×600.)

RADIOLOGICAL FINDINGS

Radiographs did not show any appreciable changes, but in two children who were treated with plaster splints shortly before radiological examination there were indications of a slight decalcification. In children old enough to have a centre of ossification in the patella this was somewhat smaller than on the normal side. The patella was usually situated rather higher than on the healthy side (Figs. 3 and 4).

ELECTROMYOGRAPHIC FINDINGS

In most children electromyography was done. The results were generally similar: the rectus femoris and vastus intermedius did not show any activity; the vastus medialis and lateralis showed a decrease of spikes to a transient state (the action potential activity was mono- or biphasic), a decrease of spikes and a reduction of frequency. In the knee flexors the pattern was either normal or showed a slight lowering of the peaks with sometimes a decrease

of potentials. The peaks were however of normal shape. (This investigation was carried out by Dr I. Lesný and Dr Vl. Janda in the laboratory of our clinic.)

Chronaximetric investigation was carried out in three children. The results were practically within normal limits. (This investigation was carried out by Dr I. Lesný.) During routine electrodiagnostic investigation the reaction of degeneration was never found.

FINDINGS AT OPERATION

Because of the failure of conservative treatment operation was advised in all cases. Ten children were operated on at the Second Orthopaedic Clinic and one was operated on elsewhere. The last child returned to her own country and nothing is known of her later course.

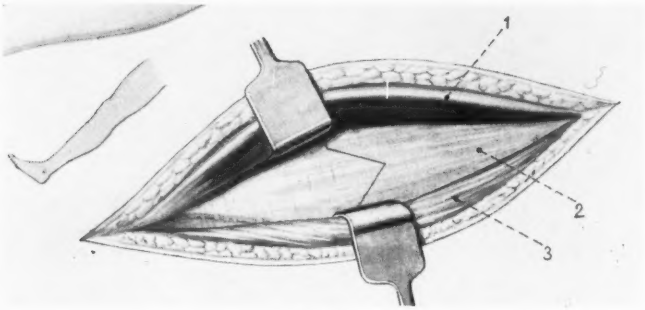


FIG. 7
Incision over medial border of rectus femoris. The vastus intermedius displayed and the line of division of its fascia shown (1—rectus femoris; 2—vastus intermedius; 3—vastus lateralis).

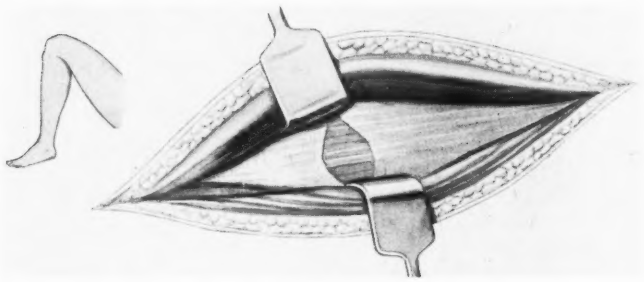


FIG. 8
Gaping of fascial incision on bending of the knee.

At operation the subcutaneous connective tissue overlying the extensor apparatus of the knee was always found to be penetrated by rather rigid fibres running vertically from the skin to the fascia, while the subcutaneous layer of fat was always rather thin. There were no very obvious changes in the muscles except perhaps a smaller volume and a higher rigidity on passive stretching. The muscles were stimulated by galvanic and faradic current; the contractions were small, slow, or completely absent (Cases 2, 3, 5, 7).

In our first cases we used the operation described by Bennett (1922), sometimes with the modification introduced by Ramadier and Lacheretz (1953). It was however seen that elongation of the contracted fascia and of the vastus intermedius was quite sufficient to permit full

flexion. In later cases therefore we limited the intervention to this procedure (Figs. 7 and 8). In some cases where the patella was markedly higher than normal the ligamentum patellae was plicated.

HISTOLOGICAL FINDINGS

Histological examination of pieces of muscle was carried out by Dr V. J. Fischer of the Institute of Pathological Anatomy (Director Professor V. Jedlička). He reports: "The diameter of muscle fibres is considerably narrowed and in many places they completely disappear.

FIG. 9

FIG. 10

FIG. 11

FIGS. 9 TO 11
Showing range of knee flexion in three children after operation.
Figure 9—Case 3. Girl aged eleven. Two years after operation.
Figure 10—Case 9. Girl aged six. Two years after operation.
Figure 11—Case 2. Boy aged five. Three years after operation.

The striping is slightly effaced. On transverse sections it can be seen that all muscle fibres show a decrease of their width and that a considerable part of all fibres of the quadriceps muscle, especially the rectus femoris and vastus intermedius, are replaced by fat. Subsarcolemmal nuclei are, in some places, activated, and endomysial nuclei increased (Figs. 5 and 6). In some places there are clusters of myophages. There is a slight increase of interstitial fat. Muscle bundles are moderately innervated, the termination of the nerve fibres giving the

impression that in some places they end blindly. This blind ending is not observed in all cases. From all investigations it is evident that there is a primary muscular disturbance. The changes in the nerves are not marked and seem to be secondary. The blind termination of nerve fibres found on histology is not constant."

The histological picture is reminiscent of that in arthromyodysplasia (amyoplasie congénitale, arthrogryposis) as reported by Poli (1929), Rocher (1933), Potel (Wiemuth 1901), Sheldon (1932), Middleton (De Palma 1954) and others.

AFTER-TREATMENT AND RESULTS

The knees were immobilised in a position of 90 degrees of flexion. Full flexion was not used because of the possibility that this might cause difficulty in re-educating the quadriceps. In all children the range of flexion of 90 degrees was maintained, though there was never even a slight improvement later (Figs. 9 to 11). The range of active flexion was nearly always the same as that of passive flexion. Knee movement up to 90 degrees is sufficient for all important movements and eventually mobility can be still more improved by a further operation in later years.

DISCUSSION

The clinical picture described resembles superficially an incomplete syndrome of arthromyodysplasia (arthrogryposis). In this condition only one part of an extremity may be affected (Cuilleret 1937), but it persists without any change from birth, and may be sometimes improved by treatment (Rocher 1933). In our cases, however, the condition first appeared in children at the age of one or more years and steadily led to increasing restriction of movement in spite of conservative treatment.

Gradual restriction of mobility due to a fibrous degeneration of certain muscles in childhood is known to occur in several syndromes: 1) Some cases of so-called congenital dislocation of the patella becoming manifest in later years only (Payr 1934, Cuilleret 1937, Leveuf and Pais 1946). Three such cases have been studied in this clinic. 2) Camptodactylia, occurring especially in girls, is often of familial incidence and is usually manifested after the tenth year of life and during puberty (Rocher and Roudil 1934, Aurousseau 1937). 3) The so-called Pouce bot flexus. 4) Some cases of digitus quintus supraductus. 5) Some cases of pectus excavatum, manifested as late as at puberty (Hněvkovský). The muscular origin of this deformity was recognised by Lester (1950).

From these analogies we may presume that in our cases there was a muscular dysplasia of congenital origin of the vastus intermedius and rectus femoris muscles, manifesting itself in later life only, progressive and not responding to conservative methods of treatment. The progressive nature of the condition is also shown by histological findings such as activation of subsarcolemmal nuclei and the presence of myophages. The congenital nature of the condition is suggested by the radiological findings of deviations and hypoplasia of the patella. The changes in the subcutaneous tissue found at operation are analogous with findings in congenital anomalies of similar character (Rocher 1933).

Fibrosis of a part of the quadriceps muscle may be the cause of different clinical conditions, according to the part of the affected muscle. If the vastus lateralis muscle is involved a lateral luxation of the patella occurs; if the middle part of the quadriceps muscle is involved it results in a " pseudoluxation "—the condition described here.

As far as I am aware, no case of deviation of the patella in a medial direction has, as yet, been described. Perhaps involvement of the ilio-tibial tract is necessary for the production of deviation, and clearly this tract cannot exert any influence on the medial side of the thigh.

As far as I am acquainted with the literature no similar case of progressive limitation of flexion of the knee caused by congenital myodysplasia has been described. Domanský (1932) described the case of a man aged forty-nine who, from childhood, had had restriction of

movement of one knee. The radiograph showed a well preserved joint space, the patella being smaller and situated rather higher than that of the other knee. Because of the limited mobility of the knee this case cannot be classified as a simple patella alta (Jansen 1929). Another similar case of congenital ankylosis with patella alta in a child of nine was published by Lombard (1934); because the operative lengthening of the quadriceps tendon was successful the author accepts the opinion of Potel (1897) that the ankylosis is of muscular origin. It is possible that more similar cases could be found but under some other diagnosis.

As to etiology, the author's view is that late disturbance of ontogenesis of the affected muscles—a dynamically active tissue—may be presumed. A congenital dysplasia of any tissue can be of varying degree and may be clinically manifested in the growth of the embryo, the foetus or the child.

SUMMARY

1. Twelve children are described in whom gradual limitation of knee flexion developed, without previous injury or inflammation.
2. Clinical and histological investigation showed progressive fibrous degeneration of the vastus intermedius and rectus femoris.
3. As the patella showed signs of retarded development a particular form of myodysplasia, probably congenital, may be supposed.
4. Conservative treatment always failed to stop the progress of this disease.
5. Surgical treatment (Bennett's method or an elongation of the ligamental apparatus of the intermedius muscle) was carried out in ten cases and gave good permanent results. Flexion remained possible only to the extent obtained at operation; rehabilitation gave no further improvement.
6. In the opinion of the author, these cases occur more often than is generally believed, but they are recorded under a different diagnosis and consequently are not submitted to rational treatment.

REFERENCES

AUROUSSEAU, L. (1937): In *Ombrédanne-Mathieu: Traité de Chirurgie Orthopédique*. Tome 3, p. 2,682. Paris: Masson et Cie.

BENNETT, G. E. (1922): Lengthening of the Quadriceps Tendon. *Journal of Bone and Joint Surgery*, **4,** 279.

COMBY, J. (1933): L'amyoplasie congénitale ou rigidité articulaire multiple congénitale. *Archives de Médecine des Enfants*, **36,** 171.

CUILLERET, F. D. (1937): In *Ombrédanne-Mathieu: Traité de Chirurgie Orthopédique*. Tome 1, p. 543. Paris: Masson et Cie.

DE PALMA, A. F. (1954): *Diseases of the Knee*. Philadelphia, London, Montreal: J. B. Lippincott Company.

DOMANSKÝ, K. (1932): O jednom případě vrozeného vymknutí čéšky směrem nahoru. *Časopis Lékařů Českých*, **71,** 1,292.

HNĚVKOVSKÝ, O., and EIS, E. (1956): Patella Alta Congenita Tarda. *Universitas Carolina Medica*, **2,** 567.

JANSEN, M. (1929): Genu impressum und Patella alta. *Zeitschrift für Orthopädische Chirurgie*, **5,** 314.

LESTER, C. W. (1950): Funnel Chest: its Cause, Effects, and Treatment. *Journal of Pediatrics*, **37,** 224.

LEVEUF, J., and PAIS, C. (1946): Les dislocations congénitales du genou. *Revue d'Orthopédie*, **32,** 313.

LOMBARD, P. (1934): Raideur congénitale du genou par brièveté du quadriceps. Allongement du tendon quadricipital et des ailerons rotuliens. *Revue d'Orthopaedie*, **21,** 127.

MIDDLETON, D. S.: Cited by De Palma (1954).

PAYR, E. (1934): Über die Behandlung der gewohnheitsmässigen oder "rückfälligen" Verrenkung der Kniescheibe. *Zentralblatt für Chirurgie*, **61,** 1,554.

POLI, A. (1929): Contributo allo studio delle assenze muscolari congenite. *Archivio di Ortopedia*, **45,** 775.

POTEL (1897): Étude sur les malformations congénitales du genou. Lille: L. Danel. (Cited by Wiemuth).

RAMADIER, J. O., and LACHERETZ, M. (1953): Traitement des ankyloses et des raideurs en extension du genou. *Revue de Chirurgie Orthopédique*, **39,** 450.

ROCHER, H. L. (1933): Les raideurs articulaires congénitales multiples. *Deuxième Congrès International de Chirurgie Orthopédique Londres*, 19–22 *juillet* 1933. Procès-verbaux, p. 570. Bruxelles: Imprimerie Lielens.

ROCHER, H. L., and ROUDIL, G. (1934): Deux observations de pouces bots flexus. *Revue d'Orthopédie*, **21,** 37.

SHELDON, W. (1932): Amyoplasia Congenita. *Archives of Disease in Childhood*, **7,** 117.

WIEMUTH (1901): Die habituellen Verrenkungen der Kniescheibe. *Deutsche Zeitschrift für Chirurgie*, **61,** 127.

VASTUS INTERMEDIUS CONTRACTURE IN EARLY CHILDHOOD

Case Report in Identical Twins

T. J. FAIRBANK and A. M. BARRETT, CAMBRIDGE, ENGLAND

Inquiry among paediatricians and orthopaedic surgeons in this country has failed to disclose any knowledge of the condition described by Hněvkovský (1961), but the twin girls reported here show typical examples. Anna and Mary W. were born in June 1956, nine weeks prematurely, the only children of a Polish father and English mother both highly intelligent and normal in every respect. The father has one sister with three normal sons, the mother one brother with three children, one of whom is a Mongol. No family history of extension contracture of the knees could be elicited.

Blood grouping confirms that the twins are identical. They were slow in developing ability to sit, stand and walk, to handle toys and to talk. Walking was first possible at two years. Because of this delayed development they were seen in September 1957 by a paediatrician who concluded that they were decidedly retarded, with a development quotient of about 70 per cent. Both were at that time anaemic, but the anaemia responded rapidly to iron therapy. They were kept under observation until July 1959; no cause was found for their delayed development or any abnormality discovered in the legs or central nervous system. Both twins were, however, still " wet " both day and night.

In December 1959, at the age of three and a half years, Anna was first seen in the orthopaedic department because the mother had noticed for the previous month that her right knee would not flex fully and that she was unable to kneel like her sister. There was no pain or restriction of other activities. There was a history of a minor injury to the knee during the previous summer which had recovered rapidly.

She was a lively child (Fig. 1) with a large brachycephalic skull, flattened posteriorly, some hypertelorism but otherwise no clinical abnormality except in the right knee, where flexion was limited to 90 degrees (Fig. 2). This limitation was not affected by the position of the hip. Flexing the knee to the end point produced a sensation of slightly springy mechanical block; forcing flexion was painless. The knee jerks were equal and normal and there was no muscle wasting: quadriceps power appeared normal. The only other abnormality detected was some restriction of antero-posterior glide of tibia on femur when the knee was flexed, and mild genu valgum. Radiographs of the knees were normal, but only the right patella showed a centre of ossification.

By April 1960 the condition of the right knee was unchanged but the left knee had lost 20 degrees of flexion, and both knees of her sister Mary had developed the same loss of range.

In May Anna was admitted to hospital. She weighed 32 pounds and her height was 39 inches. The circumference of the skull was 19½ inches. A skeletal survey revealed no abnormality apart from the shape of the skull and the fact that ossification of the carpus was slightly advanced. Blood count and blood chemistry were within normal limits. The electrophoretic pattern of the serum proteins was normal and Wassermann reaction was negative. Electromyography and strength-duration curves for each of the four heads of the quadriceps revealed no abnormality.

Findings at operation—At the end of May the lower quarter of the right quadriceps was explored. The subcutaneous tissue over the quadriceps tendon was perhaps a little indurated, fibrous and more firmly attached to the underlying tissue than elsewhere. The medial and lateral vasti were separated from the rectus tendon by sharp dissection and the rectus was

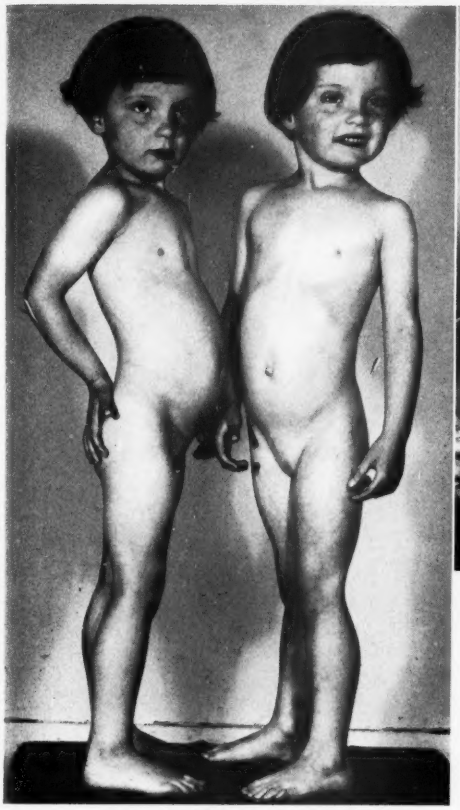


FIG. 1

FIG. 2

FIGS. 1 AND 2
Figure 1—Mary (*on the left*) and Anna (*on the right*), May 1960. Figure 2—Anna's limitation of knee flexion, May 1960.

lifted from the underlying intermedius. Muscle and tendon of all four heads appeared macroscopically normal. The range of flexion was not improved under anaesthesia. With the knee passively flexed to the maximum the rectus and vasti medialis and lateralis were not tense, but the tendon of intermedius was bar-tight. When this tendon was divided flexion through a further 40 degrees was immediately possible without force; full flexion was possible with moderate force but was "springy." One inch of the intermedius tendon was excised, specimens of all four muscle heads were taken, and the medial and lateral vasti sutured back to the sides of the rectus tendon. The knee was kept flexed 40 degrees past the right angle in plaster-of-Paris until the sutures were removed on the tenth day.

Progress—Recovery was uneventful. Within three months she could walk six miles and run normally. There has been no loss of the flexion gained at

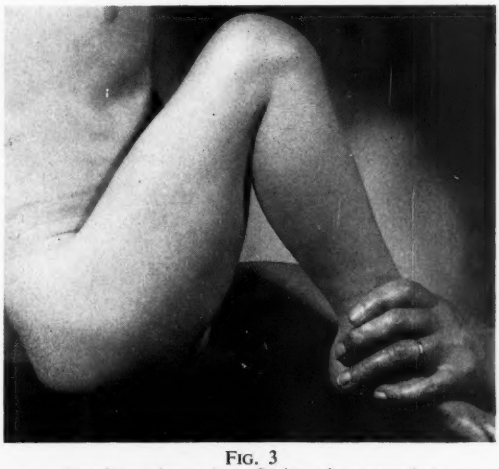


FIG. 3
Anna's maximum knee flexion after operation.

FIG. 4

FIG. 5

Figure 4—Vastus intermedius, showing excess of adipose tissue and deficiency of muscle. Note fat separating individual muscle fibres. (Weigert's elastic stain and Curtis's ponceau S, ×110.) Fibrous tissue appears black.
Figure 5—Vastus intermedius, showing excess of collagen (black). The rather thin, wavy black fibres at and to the left of centre are striated collagen fibres as shown in Figures 13 and 14, but the striations are invisible in this low-power view. (Weigert's elastic stain and Curtis's ponceau S, ×110.)

FIG. 6

Rectus femoris: normal. (Weigert's elastic stain and Curtis's ponceau S, ×110.) Fibrous tissue appears black.

operation (Fig. 3) or further deterioration in the other knee. During the brief period of observation the condition of Mary's knees did not deteriorate.

HISTOLOGICAL FINDINGS

Samples removed at operation from the following sites were examined histologically: rectus femoris, vastus lateralis, vastus medialis, fascia superficial to the vastus intermedius, and the tendon and adjacent muscle of the vastus intermedius. The samples were fixed in Heidenhain's " Susa " fixative, which is the fixative used by us as routine for small biopsy specimens; they were embedded in paraffin, and sections were stained by Ehrlich's haematoxylin and eosin, Weigert's elastic stain and Curtis's ponceau S, Mallory's phosphotungstic acid haematoxylin and McFarlane's picro-Mallory method. Longitudinal and transverse portions of the muscle samples were embedded and sectioned separately.

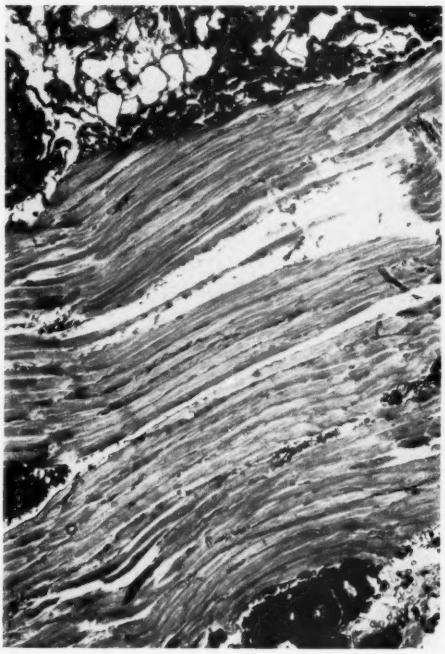


FIG. 7

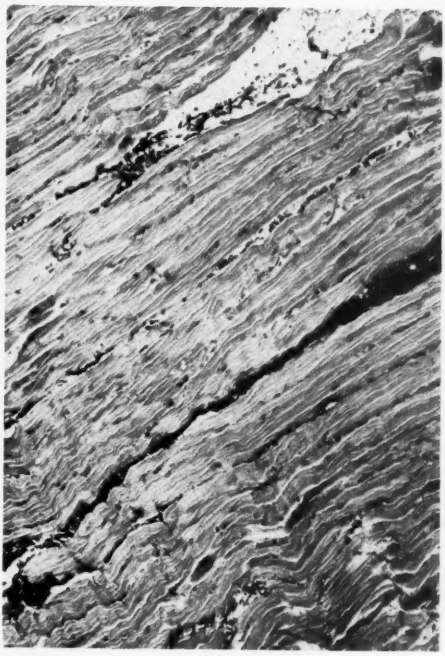


FIG. 8

Figure 7—Vastus medialis. Note the fat at the top of the section. (Weigert's elastic stain and Curtis's ponceau S, ×110.) Figure 8—Vastus medialis. (Weigert's elastic stain and Curtis's ponceau S, ×110.)

The only definite abnormalities observed were in the vastus intermedius muscle and the adjacent fascia. In this muscle there was an excess of adipose and fibrous tissues and a deficiency of muscular tissue in marked contrast to the normal appearance of the rectus femoris (Figs. 4 to 6). The fat was present not only between the large bundles of muscle fibres, but within them, separating individual muscle fibres (Fig. 4). In contrast to this, where there was any fat in the sections of the other muscles (Fig. 7) it was restricted to the larger connective tissue septa, and there was much less of it. Figure 7 was deliberately chosen to include such adipose tissue, and Figure 8 is more representative of the same muscle (vastus medialis).

Some parts of the residual muscle in the vastus intermedius appeared normal, but most of the muscle fibres were smaller than those in the other muscles and nuclei appeared more

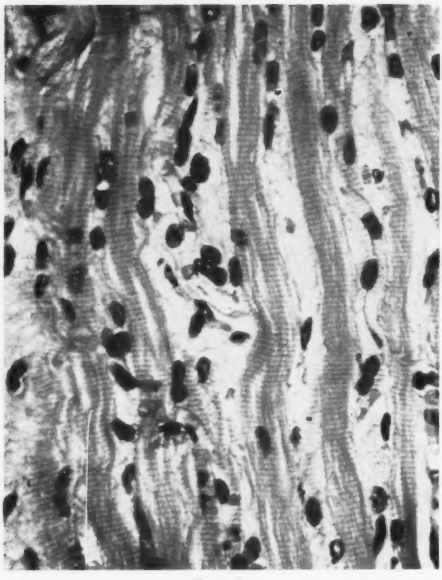


FIG. 9

FIG. 10

Contrasting appearances of vastus intermedius and rectus femoris. Figure 9—Vastus intermedius; longitudinal section. (Ehrlich's haematoxylin and eosin, ×420.) Figure 10—Rectus femoris; longitudinal section. (Ehrlich's haematoxylin and eosin, ×420.)

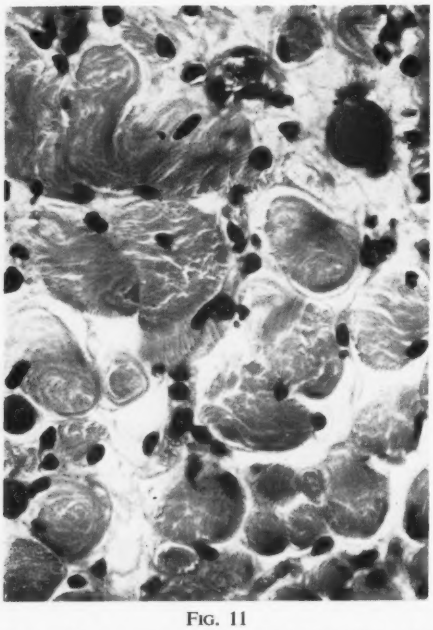


FIG. 11

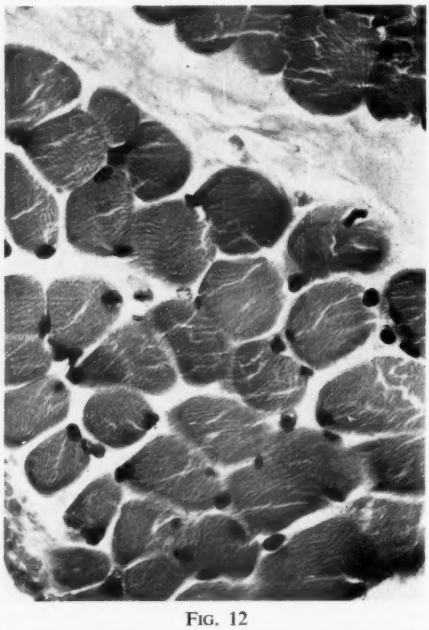


FIG. 12

Contrasting appearances of vastus intermedius and vastus lateralis. Figure 11—Vastus intermedius; transverse section. (Ehrlich's haematoxylin and eosin, ×420.) Figure 12—Vastus lateralis; transverse section. (Ehrlich's haematoxylin and eosin, ×420.)

abundant (Figs. 9 and 10). Most of the excess nuclei were nuclei of endomysial cells, not of muscle fibres. The decrease in size of the muscle fibres would be expected to bring the muscle fibre nuclei closer together, and allowing for this there was not deemed to be evidence of actual increase in the number of nuclei in the muscle fibres. Occasional single fibres here and there were much more atrophic than the rest, but there were no groups of very small fibres

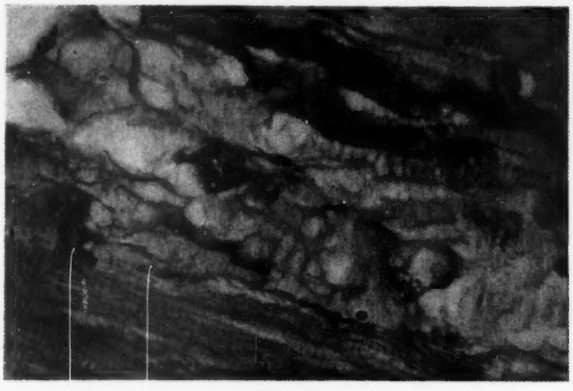


FIG. 13

Vastus intermedius. Striated collagen fibres in a section stained by Curtis's ponceau S variant of van Gieson's stain. Striated collagen fibres (red) at top left; striated muscle fibres (yellow) at bottom right. (×600.)

FIG. 14

Vastus intermedius. A striated collagen fibre at a higher magnification than in Figure 13. (Curtis's ponceau S variant of van Gieson's stain, ×1,100.)

such as are often seen in paralytic atrophy, and none of the abnormally large fibres usual in muscular dystrophies. The muscle fibres were abnormally variable in size, structure and staining reactions, but this increased variability was of relatively slight degree. It was most obvious in transverse sections (Figs. 11 and 12), and in some of the fibres which were apparently cut in a true transverse plane the fibrillae appeared to be running obliquely. The transverse striations were very well preserved (Fig. 9).

The sections of the vastus intermedius and the vastus medialis included muscle spindles and small nerves. In the vastus medialis these appeared normal; in the vastus intermedius the number of nerve fibres appeared small compared to the size of the nerve and the myelin sheath appeared swollen, but neither of these features was definitely outside the range of normal variations.

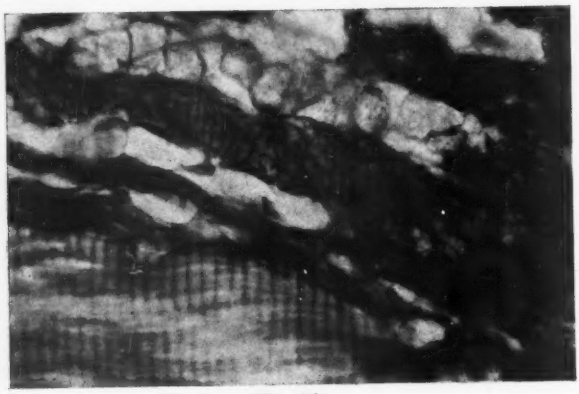


FIG. 15

Vastus intermedius. A striated collagen fibre (blue) and a striated muscle fibre (red) in a section stained by McFarlane's picro-Mallory method. (×1,100.)

FIG. 16

Vastus intermedius. A striated collagen fibre (pink) in a section stained by Mallory's phosphotungstic acid haematoxylin. Part of a muscle fibre (purple-blue) is included at the bottom edge of the photograph on the right. (×1,100.)

The most remarkable histological feature of the vastus intermedius was the presence of fibres with the staining reactions of collagen but with transverse striations similar to those of muscle fibres (Figs. 13 to 16). These fibres were quite inconspicuous in sections stained with haematoxylin and eosin in which they were very pale pink, but with Curtis's ponceau S stain (Figs. 13 and 14), McFarlane's picro-Mallory method (Fig. 15) and Mallory's phosphotungstic acid haematoxylin (Fig. 16) their staining reactions were identical with those of collagen fibres, and quite unlike those of muscle. No fibres intermediate between these striated collagen

fibres and muscle fibres were seen. Measurements showed that the spacing of the striations in the collagen fibres was very nearly the same as that of the striations of the adjacent muscle fibres. The thin wavy black fibres at and to the left of centre of Figure 5 are in fact the striated collagen fibres shown at high magnification in Figures 13 and 14, and they can be seen to have the same direction as the adjacent muscle fibres, but to be much thinner than the muscle fibres.

In contrast to the sections of the vastus intermedius, those of the rectus femoris, vastus medialis and vastus lateralis appeared almost or quite normal. The rectus was certainly normal; very slight changes of the kind seen in the vastus intermedius may have been present in vastus medialis and vastus lateralis, but if so they were too slight to be recognised with certainty. No striated collagen fibres were seen except in the vastus intermedius.

DISCUSSION

The histological changes are very similar to those described by Hněvkovský, although in the present case the changes were practically restricted to the vastus intermedius; and whereas in Hněvkovský's cases the striations of the muscle fibres were slightly effaced, in the present case they were well preserved.

Striated collagen fibres were not described by Hněvkovský, and we ourselves have never seen them before, nor have we come across any description of them. The fact that the spacing of the striations was almost the same as that of the striations of the muscle fibres, and that the direction of the collagen fibres was the same as that of the adjacent muscle fibres, at once suggested that muscle fibres might have been converted into collagen without losing their striations. This would be a most remarkable transformation. Although Krösing (1892), Durante (1902), Tower (1935) and Altschul (1942) described the transformation of muscle fibres into fibrous tissue or adipose tissue in muscular atrophy resulting from denervation, loss of transverse striations was described as an early change, and none of these workers reported the persistence of transverse striations in the collagen fibres which they thought resulted from the fibrotic de-differentiation of muscle. Bowden and Gutmann (1944) and Adams, Denny-Brown and Pearson (1953) failed to confirm the transformation of muscle fibres into collagen fibres. We do not consider that our observations provide strong enough evidence to justify a claim that a transformation as remarkable as the conversion of muscle fibres into striated collagen fibres had occurred. Before making such a claim, we would wish to see intermediate stages between muscle and collagen, and in our sections there are none. We would also wish to see striated collagen fibres in tissues fixed in fixatives other than "Susa." "Susa" is not a good fixative for connective tissues, because collagen is partially acid-soluble, and acetic acid and trichloracetic acid, which "Susa" contains, cause an alteration in the appearance of collagen fibres, so that they become finely vacuolated or foamy under a high magnification. This foamy appearance is conspicuous in many of the collagen fibres in our sections, and possibly the appearance of striation had resulted from some kind of alignment of the vacuoles: there are apparent transitional stages between striated fibres and those which were merely foamy. But we cannot explain why the striated fibres were seen only in the vastus intermedius sections, or why we have never seen them before, although we have used "Susa" as a routine fixative for many years.

The examination of collagen fibrils with the electron microscope constantly reveals transverse striations, but these are very much finer than the striations in our striated collagen fibres: the periodicity of the normal striations is 640 Å and they are quite invisible with even the highest magnification of the ordinary light microscope.

Replacement of muscle fibres by fibrous tissue and adipose tissue are non-specific changes which may occur in the late stages of various atrophic or degenerative conditions. In our case they provide evidence that the vastus intermedius muscle had long been abnormal, probably since the time of birth or earlier, but they do not elucidate the nature of the disorder; neither

do the relatively slight abnormalities in the residual muscle. There are features in common with those described by Gilmour (1946) and others in amyoplasia congenita (arthrogryposis), but these features are also seen in other disorders, and in our opinion the resemblance is not close enough to be of much significance. Nor is the histological picture very much like that seen in paralytic atrophy or in most cases of muscular dystrophy, although some form of myopathy remains a possibility. Contracture of the affected muscles is uncommon in muscular dystrophy, and when it occurs is usually a late change in severely afflicted muscles or of the stronger muscles opposed by weakened groups. On the whole, vastus intermedius contracture seems to us to have most in common with the contracture of the sternomastoid muscle which results in " congenital " torticollis.

The occurrence of this condition in identical twins is suggestive of a genetic origin. If indeed it is such a rarity in this country the fact that the father is Polish, when viewed in the light of Hněvkovský's paper, suggests the possibility that we are concerned with a disease peculiar to Eastern Europe.

Our thanks are due to Dr M. Yealland and Dr W. A. Fell for performing the electromyography and plotting the strength-duration curves, to Mr S. W. Patman for the black and white photomicrographs, and to Mr R. J. Flemans for those in colour.

REFERENCES

ADAMS, R. D., DENNY-BROWN, D., and PEARSON, C. M. (1953): *Diseases of Muscle.* New York: Paul B. Hoeber Inc.

ALTSCHUL, R. (1942): Atrophy, Degeneration and Metaplasia in Denervated Skeletal Muscle. *Archives of Pathology*, **34**, 982.

BOWDEN, R. E. M., and GUTMANN, E. (1944): Denervation and Re-innervation of Human Voluntary Muscle. *Brain*, **67**, 273.

DURANTE, G. (1902): Du processus histologique de l'atrophie musculaire. *Archives de Médecine Expérimentale et d'Anatomie Pathologique*, **14**, 658.

GILMOUR, J. R. (1946): Amyoplasia Congenita. *Journal of Pathology and Bacteriology*, **58**, 675.

HNĚVKOVSKÝ, O. (1961): Progressive Fibrosis of the Vastus Intermedius Muscle in Children. *Journal of Bone and Joint Surgery*, **43–B**, 318.

KRÖSING, R. (1892): Ueber die Rückbildung und Entwickelung der quergestreiften Muskelfasern. *Archiv für Pathologische Anatomie und Physiologie*, **128**, 445.

TOWER, S. S. (1935): Atrophy and Degeneration in Skeletal Muscle. *American Journal of Anatomy*, **56**, 1.

A CASE OF MELORHEOSTOSIS WITH CUTANEOUS HAEMANGIOMA AND LYMPHATIC VESICLES

R. HALL, NEWCASTLE-UPON-TYNE, ENGLAND

Melorheostosis is a rare benign sclerotic condition of bone first described by Léri and Joanny in 1922. It is recognised by the characteristic radiological appearance of the sclerosis, which resembles the flow of candle grease down a bone. The lesion usually affects the bones of a single limb, but cases of wider distribution have been described. Elkeles (1954) reported a case in which all four limbs and the pelvis were involved. Pain is often present in the affected area and joint movement may be limited.

CASE REPORT

A fifteen-year-old girl complained of painful swelling of the right foot for two years, slowly progressing to involve the whole limb. There was a diffuse " port wine " naevus on the right side of the lower chest and abdomen. The right hand was larger than the left and showed spindling of the fourth and fifth fingers. The right leg was enlarged up to the groin and

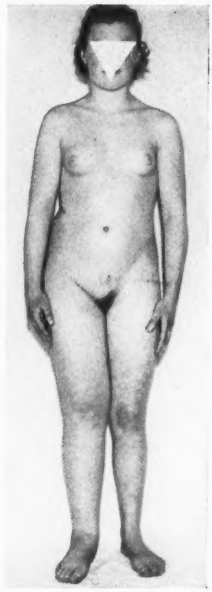


FIG. 1

FIG. 2

Figure 1—"Port wine" naevus, enlarged right lower limb, with lymphatic vesicles in groin. Figure 2—Lymphatic vesicles in groin.

there was pitting oedema up to the knee (Fig. 1). There were multiple lymphatic vesicles in the right groin containing milky fluid, but no evidence of scleroderma (Fig. 2). Deep induration could be felt in the right femoral triangle. The right lower limb was one and a half inches (four centimetres) longer and the thigh and calf circumferences were two inches (five centimetres) and two and a half inches (six centimetres) respectively greater than the corresponding measurements on the left side. No thrill could be felt and no bruit heard over the right leg.

FIG. 3
Radiograph of hands showing bone sclerosis on right side.

FIG. 4
Radiograph of right half of pelvis and right femur showing bone sclerosis and ossification in adductor region of right thigh.

Radiographic changes characteristic of melorheostosis, confined to the right side, were found in the following bones: styloid process of ulna, triquetrum, hamate, capitate, medial three metacarpals, all phalanges of fifth finger, proximal phalanx of fourth finger (Fig. 3), pubis, femur, tibia, talus, navicular, first and second cuneiforms and metatarsals, and the proximal phalanx of the hallux. There was ossification in the adductor region of the thigh (Fig. 4). The increased length of the right lower limb was confirmed by radiological measurements. The chest radiograph was normal. The serum calcium was 9·6 milligrams per 100 millilitres, serum phosphate 3·2 milligrams per 100 millilitres, and the alkaline phosphatase 7·9 units (Jenner and Kay). The haemoglobin was 94 per cent, colour index 0·94. The electrocardiograph was normal. No evidence of arteriovenous fistulae could be demonstrated by an aortograph or by plethysmography, which showed calf blood flows of 3·5 millilitres per 100 g. tissue per minute on the right side and 7·4 millilitres on the left.

No specific treatment is possible and the parents and patient were reassured of the benign nature of the condition. The sole of the left shoe was thickened and the swelling of the right leg controlled by a full-length elastic stocking.

DISCUSSION

The etiology of the condition is unknown, but it is probably a congenital disorder of bone as suggested by Léri and Joanny (1922). The association of the condition with neurofibromatosis has been described by McCarroll (1950) and by Lièvre (1953). A cutaneous haemangioma was present in one of the cases described by McCarroll. It is of interest that both lymphatic vesicles and cutaneous haemangioma have been described in cases of neurofibromatosis. Arteriovenous aneurysms are one cause of giant limb and occurred in the case of melorheostosis described by Murray (1951). No definite evidence of arteriovenous fistulae could be found in this case but it remains possible that multiple small congenital arteriovenous communications were present in the limb.

Subcutaneous bony deposits like that seen in this case have been reported on several occasions, for instance by Kraft (1932).

SUMMARY

A case of melorheostosis is described in which more than one limb was affected. It was associated with increased length of the right leg, lymphatic vesicles in the right groin, ossification in the subcutaneous tissues of the right thigh and a cutaneous haemangioma of the right side of the trunk.

I wish to thank Mr J. C. Clark for permission to publish the case, Dr Irwin who suggested the diagnosis and Professor A. G. R. Lowdon for his advice.

REFERENCES

ELKELES, A. (1954): Melorheostosis Involving All Limbs, Pelvis and Sacrum: X-ray. *Transactions of the Medical Society of London*, **70,** 204.

KRAFT, E. (1932): Melorheostosis Léri: A Flowing Hyperostosis of a Single Extremity. *Journal of the American Medical Association*, **98,** 705.

LÉRI, A., and JOANNY (1922): Une affection non décrite des os; hyperostose " en coulée " sur toute la longeur d'un membre ou " mélorhéostose." *Bulletins et Mémoires de la Société Médicale des Hôpitaux de Paris*, Troisième Série, **46,** 1,141.

LIÈVRE, J. A. (1953): Mélorhéostose. Observation et discussion pathogénique. *Revue du Rhumatisme*, **20,** 788.

MCCARROLL, H. R. (1950): Clinical Manifestations of Congenital Neurofibromatosis. *Journal of Bone and Joint Surgery*, **32–A,** 601.

MURRAY, R. O. (1951): Melorheostosis Associated with Congenital Arteriovenous Aneurysms. *Proceedings of the Royal Society of Medicine* (Section of Orthopaedics), **44,** 473.

PRIMARY OSTEOSARCOMA OF THE PATELLA

MARTIN A. GOODWIN, DARLINGTON, ENGLAND

From the Darlington Memorial Hospital

Primary tumours of the patella are rare. Benign growths, exostoses and chondromata are even less common than malignant ones. Giant-cell tumour (osteoclastoma), a benign lesion with malignant potentialities, appears to be the most frequent tumour at this site and, in all, possibly a score of cases have been reported.

Osteosarcoma of the patella is certainly rare. Christensen (1925), reviewing 1,000 cases of bone tumours collected from the Codman Registry and from the literature, found only one case of sarcoma of the patella, and Hambly (1959) put the number of acceptable cases of sarcoma of the patella in the literature at only thirteen. The present case of primary osteosarcoma of the patella with lung metastases therefore seems worth recording, and the opportunity has been taken to give a brief review of the literature.

CASE REPORT

A man of twenty-four slipped and hurt his left knee in January 1951. He slipped again in March of that year and was then admitted to a hospital in the south of England, complaining of pain in the left knee. The clinical appearance suggested infective arthritis, and arthrotomy revealed what was described as thick pus and granulation tissue. A long leg plaster was applied and a course of penicillin was given. In April 1951 he was transferred as an out-patient to the orthopaedic clinic of the General Infirmary at Leeds, where I saw him for the first time.

The plaster was removed in May 1951 and mobilisation of the knee commenced. The knee was swollen and painful, particularly over the patella. A radiograph showed generalised osteoporosis with erosion of the inferior pole of the patella (Fig. 1). Arthritis was suspected and his admission to hospital was arranged. The erythrocyte sedimentation rate was three millimetres in the first hour and the white cell count was 8,100 per cubic millimetre. A radiograph of the chest two months later showed a normal appearance.

Operation—The knee was opened through a medial parapatellar incision. The patella was soft and eroded, the appearances suggesting osteomyelitis. There was much granulation tissue in the joint, and the suprapatellar pouch was thickened. The patella and suprapatellar pouch were excised.

Histological examination (Professor D. H. Collins)—Marrow spaces in various parts of the patella were filled by a highly cellular tumour which appeared to be an intramedullary spindle-celled sarcoma (Fig. 3). There was no evidence of infection. The suprapatellar pouch was extensively infiltrated with sarcomatous tissue composed of the same type of spindle cells as were seen within the patella.

Further treatment and progress—A mid-thigh amputation was performed two months after the first operation. Six months later radiographs of the chest showed a large rounded opacity in the left lower zone, with two other smaller opacities seen only in the lateral film (Fig. 2). The patient later developed acute retention of urine, and he eventually died from metastases.

DISCUSSION

The number of published cases of primary osteosarcoma of the patella has been variously estimated. The earliest reviews of the literature are by Kienböck (1924) and Cole (1925), and all the cases from these reviews are included among the list of twenty-seven cases quoted by Bowen (1943). This list includes cases in which the histological description of the tumour is

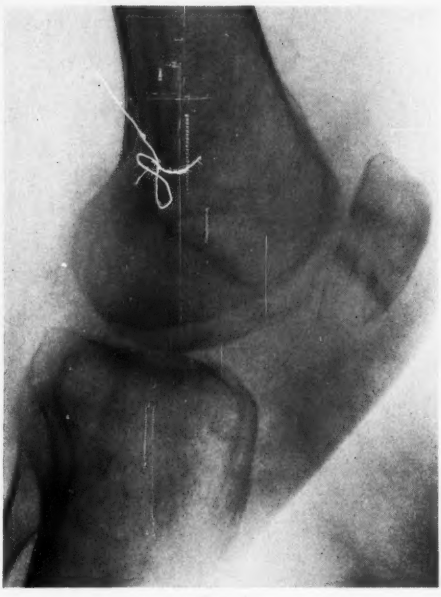


FIG. 1

FIG. 2

Figure 1—Radiograph showing the erosion of the lower pole of the patella. Figure 2—Lateral view of the chest six months after amputation of the limb, showing the large opacity in the lower zone suggestive of a secondary sarcomatous deposit.

FIG. 3

Section of the patella. The marrow spaces are filled by a highly cellular spindle-celled sarcoma. (×100.)

inadequate or absent and others in which a clear distinction is not made between osteosarcoma and so-called myeloid tumours. Blumensaat (1936) was careful to distinguish between giant-cell tumour (osteoclastoma) and true osteosarcoma, and considered that twenty-two cases of true osteosarcoma had been recorded, but he had doubts about the true nature of some of the tumours included among these twenty-two cases. Reference has already been made to Hambly's (1959) statement that only thirteen acceptable cases of sarcoma of the patella are to be found in the literature, and this includes cases described as "myeloid disease" and "myeloid sarcoma."

TABLE I

EIGHT CASES OF OSTEOSARCOMA OF THE PATELLA

Author	Clinical details	Histology
Neumann 1871	No clinical details given. Tumour excised because of ulceration	Periosteal spindle-celled sarcoma
Hayem and Graux 1874	Woman aged 22 years, lung metastases	Histology said to show sarcoma of patella
Wild 1882	Woman aged 30 years. Mid-thigh amputation. No recurrence 16 months later	Round oval- and spindle-celled sarcoma
Parker 1896	Girl aged 13½ years. Local resection. Died 7 years later from metastases	Periosteal spindle-celled sarcoma
Creite 1906	Man aged 42 years. Local removal and excision of knee joint. Died 6 months later	Spindle-celled sarcoma with giant cells
Schmidt 1907	Woman aged 45 years. Mid-thigh amputation. Well 3½ years later	Small spindle-celled sarcoma
Bianchetti 1926	Local excision. Metastases 8 months later	Small-celled sarcoma
Bellini 1934	Man aged 23 years. High amputation. Death from lung metastases	Primary endothelial sarcoma

Table I lists cases from the literature which by fairly strict criteria seem acceptable as primary osteosarcoma of the patella. Many of the cases included in previous reviews have been rejected on account of inadequate histological descriptions of the tumour, or failure to distinguish between osteosarcoma and so-called myeloid tumour. I have also excluded cases in which osteosarcoma appears to have arisen in a previously existing giant-cell tumour (the case of Bobbio (1935) and Bowen's personal case), cases of multiple bone tumours in which it could not be established that the patella was the primary site (Pillet 1931), and cases in which multiple chondrosarcomata was a possible alternative diagnosis, as in Blumensaat's personal case.

Trauma as a precursor and possibly as a causative factor is always seriously considered by continental authors. Yet considering the exposed site of the knee cap and its frequent liability to injury the incidence of malignant disease in relation to trauma is exceedingly small.

Christensen (1925) suggested that the immunity to bone tumours which the patella enjoys "is probably due to the absence of loss of growth restraint, incident to active diaphysial growth and pressure epiphysis. The fact that the patella develops from an endochondral centre, that it has a relatively short period of growth and that it is a sesamoid bone, may be of importance."

In differential diagnosis inflammatory conditions of the knee joint have to be considered. In many of the reported cases synovitis of the knee—either pyogenic or tuberculous—was the first diagnosis. Osteomyelitis was also considered, neoplasm being finally established after biopsy.

SUMMARY

1. A case of primary osteosarcoma of the patella is reported.
2. A brief review of the literature has been made.
3. In the author's opinion only nine genuine cases have been reported, including the present case.

I wish to thank Mr R. Broomhead for his permission to publish this case, Professor D. H. Collins for his help in the histological examination, Dr P. N. Coleman for his criticism, and Mr S. Taylor for the photographs.

REFERENCES

BELLINI, A. (1934): Di un sarcoma endoteliale primitivo della rotula. *Clinica Chirurgica*, **10,** 975.

BIANCHETTI, C. F. (1926): Sopra un caso di frattura spontanea di rotula affetta da sarcoma primitivo periosteo a cellule polimorfe. *Chirurgia degli Organi di Movimento*, **11,** 46.

BLUMENSAAT, C. (1936): Die Tumoren der Kniescheibe. *Ergebnisse der Chirurgie und Orthopädie*, **29,** 347.

BOBBIO, L. (1935): Sarcoma di rotula neoformata. *Bollettino e memorie della Società piemontese di Chirurgia*, **5,** 765.

BOWEN, F. H. (1943): Sarcoma of the Patella. *Journal of the Florida Medical Association*, **30,** 20.

CHRISTENSEN, F. C. (1925): Bone Tumors. *Annals of Surgery*, **81,** 1,074.

COLE, W. H. (1925): Primary Tumors of the Patella. *Journal of Bone and Joint Surgery*, **7,** 637.

CREITE (1906): Beitrag zur Pathologie der Kniescheibe. *Deutsche Zeitschrift für Chirurgie*, **83,** 179.

HAMBLY, E. (1959): Primary Tumour of the Patella. *Proceedings of the Royal Society of Medicine* (Section of Orthopaedics), **52,** 576.

HAYEM, G., and GRAUX, G. (1874): Sarcome primitif du tendon rotulien droit. *Gazette Médicale de Paris*, Série 4, **3,** 298.

KIENBÖCK, R. (1924): Über die Sarkome der Patella. *Fortschritte auf dem Gebiete der Röntgenstrahlen*, **32,** 517.

NEUMANN, E. (1871): Zur Kenntniss der zelligen Elemente der Sarcome. *Archiv der Heilkunde*, **12,** 66.

PARKER, R. W. (1887, 1896): A Case of Removal of Right Patella for Primary Sarcoma. *Transactions of the Clinical Society of London*, **20,** 254; **29,** 22.

PILLET, D. (1931): Sarkom der Kniescheibe mit Tochtergewächsen in verschiedenen Knochen, Tod durch Lungentochtergewächs des Kniescheibensarkoms—keine Unfallfolgen. *Monatsschrift für Unfallheilkunde und Versicherungsmedizin*, **38,** 311.

SCHMIDT, W. (1907): *Über die von der Patella ausgehenden Geschwülste mit besonderer Berücksichtigung des primären Sarkoms.* Leipzig: B. Georgi.

WILD, G. (1882): Zur Casuistik der Periostsarkome. *Deutsche Zeitschrift für Chirurgie*, **17,** 548.

RESULT AFTER THIRTY-ONE YEARS OF REPAIR OF TORN CRUCIATE LIGAMENTS

EDRIC F. WILSON, PLYMOUTH, DEVON

An advantage of practising in the same locality for a few decades is the opportunity sometimes afforded of following the long term results of operations, as illustrated by the following case.

In 1924 a man aged twenty sustained a complete tear of both cruciate ligaments of the knee in a severe injury at football. The resulting instability was very disabling, and such physiotherapy as was then available in a small hospital had little effect. Operative reconstruction was carried out by the method described by Smith (1918). The anterior cruciate ligament was reformed by a strip of fascia lata which was passed through drill holes in the lateral condyle of the femur and the medial condyle of the tibia. It was then sutured to the medial condyle of the femur, so re-forming the medial ligament. The posterior cruciate ligament was re-constructed by passing the tendon of semitendinosus through a drill hole in the head of the tibia.

FIG. 1

The post-operative progress was satisfactory and the patient ultimately returned to his work as a boilermaker in the naval dockyard. I did not see him again until June 1955, when he had been admitted at the age of fifty years with symptoms suggestive of carcinoma of the oesophagus. At operation a columnar-cell carcinoma of the lowest third of the oesophagus was found and oesophagectomy was performed.

While he was in hospital the knee was examined.

Since the reconstruction thirty-one years before he had had no serious trouble with the knee. Though he never again attempted to play football, he had played tennis. There was slight laxity of the medial ligament and cruciate ligaments and a little wasting of the vastus medialis. The plane of the antero-posterior movement of the flexed tibia was abnormal, being forward and medial instead of forward and lateral. Movement was full, with a little crepitus. Radiographs showed advanced osteoarthritis with cupping of the articular surfaces of the tibial condyles and calcification of the articular cartilage. The changes were more marked in the medial compartment.

The patient died in March 1956 and the knee joint was sent to the Royal College of Surgeons. I am indebted to Dr L. W. Proger, Pathology Curator to the Royal College of Surgeons of England, for the photograph of the specimen (Fig. 1) and for the catalogue description. " The right knee joint exposed from the front in the flexed position. The patella has been turned down to expose the reconstructed cruciate ligaments. The articular surfaces of the femur show a moderate degree of osteoarthritis, with patchy erosion of the cartilage and lipping of the margins."

Comment—The treatment of this injury appears to have changed in recent years, reliance being placed on rehabilitation of the muscular system of the knee. Nevertheless the result of operative reconstruction of the ligaments in this case suggests that the position might be reconsidered. It is interesting to look back at the opinions on treatment put forward many years ago. Jones and Smith (1913) wrote that " the rupture of any ligaments of the knee should be treated by absolute rest until healing is complete. This of course applies with the greatest force to rupture of the cruciate ligaments. . . . In old cases where pain, disability, and recurring effusions occur, the choice should be between an operation for suturing the crucials, a mechanical support allowing flexion, or an arthrodesis of the joint."

REFERENCES

JONES, R., and SMITH, S. Alwyn (1913): On Rupture of the Crucial Ligaments of the Knee, and on Fractures of the Spine of the Tibia. *British Journal of Surgery*, **1**, 70.

SMITH, S. Alwyn (1918): The Diagnosis and Treatment of Injuries to the Crucial Ligaments. *British Journal of Surgery*, **6**, 176.

INTRATHECAL SPINAL TUMOUR AS A CAUSE OF COCCYDYNIA

Report of a Case

R. NEVILLE HOULDING and A. T. MATHESON, BULAWAYO, SOUTHERN RHODESIA

There has been periodic dispute regarding the cause of coccydynia and its appropriate treatment. Among the causes that have been proposed are local conditions of the coccyx and sacro-coccygeal joints, median lumbar disc protrusions (Richards 1954) and small tumours of the glomus coccygeus (Klem 1948, Reuther 1938). The patient to be described presents an additional possible cause which merits consideration. A further aspect is that whereas many authors have drawn attention to the frequency with which cauda equina tumours present with backache and sciatic pain (Toumey, Poppen and Hurley 1950), no reference can be found in the recent literature to such tumours causing coccygeal pain only.

CASE REPORT

A European man of thirty-eight had suffered from coccydynia since 1950. Pain was first felt just beyond the tip of the coccyx while he was working under a motor car. Three years later the coccyx was excised in part, with relief of pain for several months.

When he was first seen by us in 1954 the sole complaint was that of coccygeal pain. Examination showed a healed and non-tender scar of the previous operation. He also had a small internal haemorrhoid which was treated by injection. Radiographs showed that the terminal segment of the coccyx had been removed. There was slight osteoarthritic spurring of the sacro-coccygeal joint. No treatment was advised.

Subsequently he sought other advice but he returned to consult us in September 1959. He reported then that Butazolidine was effective in relieving the coccygeal pain but that he was concerned about the possible cumulative effects of the drug. Examination showed slight restriction of straight leg raising. Radiographs of the lumbar spine and sacro-iliac joints were normal. The opinion was given that his pain might well arise from a recurrent small median lumbar disc protrusion but that he should continue to take Butazolidine in reasonable doses.

In February 1960 he first reported additional symptoms. The pain now extended from in front of the coccyx down the back of both legs. It was aggravated by coughing, jarring and constipation and was no longer relieved by Butazolidine. He also mentioned intermittent numbness in the three lateral toes of the right foot. Although examination again revealed no particular abnormality, it was decided that myelography should be performed.

Lumbar puncture was performed at the 3–4 interval. Although cerebro-spinal fluid did not flow freely at that level, Myodil was injected: this was found to be partly extra-thecal. One week later puncture was repeated at the lumbar 2–3 interval. Cerebro-spinal fluid escaped freely under raised pressure. It contained haemosiderin, Myodil, protein at over 100 milligrams per 100 millilitres, and no cells. Examination after injection of Myodil showed a space-occupying lesion opposite the body of the fourth lumbar vertebra (Fig. 1) suggesting a large intrathecal spinal tumour.

Operation—The lamina of the fourth lumbar vertebra and parts of the laminae of the third and fifth lumbar vertebrae were removed on both sides. Considerable fibrosis, presumably from the extrathecal injection of radio-opaque fluid, was noted about the ligamentum flavum at the 3–4 level. No other abnormality was noted extrathecally, but the thecal sac was greatly expanded in the upper half of the exposure and was not pulsating. When it was opened a large fleshy red tumour extruded itself into the wound. Above and below it was attached only to a large tortuous vein, and it was removed completely after division of the vein between ligatures.

Progress—Recovery was uneventful. The paraesthesiae in the right small toes subsided after a week, and six weeks after operation coccygeal pain had not recurred.

Histology—Sections of the tumour showed that it was a papillary ependymoma, without the myxomatous degeneration commonly found in ependymal tumours from the cauda equina (Fig. 2).

FIG. 1

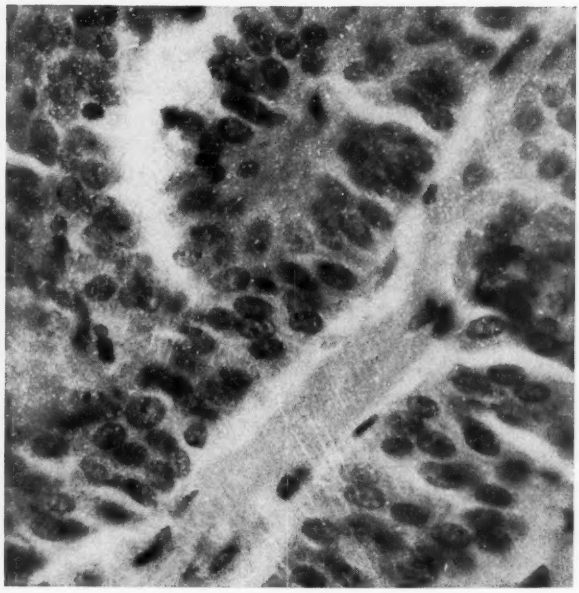


FIG. 2

Figure 1—Myelograph showing a complete block below the level of lumbar 3–4 interspace. (Some of the fluid is extrathecal.) Figure 2—Section of the tumour: the histology is that of an atypical ependymoma. (×700.)

SUMMARY

1. Coccygeal pain in a young man, persisting for ten years in all and for seven years after partial removal of the coccyx, was shown to be due to an intrathecal tumour of the cauda equina.
2. Removal of the tumour, which was found to be an ependymoma, gave complete relief.

Appreciation is expressed to Dr P. J. Barnard of Bulawayo, and to Professor Norman M. Dott and Dr A. F. J. Maloney of Edinburgh for their opinions on the histology. Our thanks are also due to Dr P. Palmer and Dr B. Tulloch, of the General Hospital, Bulawayo, for the photographs.

REFERENCES

KLEM, K. K. (1948): Glomustumor utviklet fra glomus coccygeum. *Nordisk Medicin*, **40**, 1,999.

REUTHER, T. F. (1938): The Coccygeal Glomus: a Possible Factor in Coccygodynia. *Illinois Medical Journal*, **73**, 134.

RICHARDS, H. J. (1954): Causes of Coccydynia (discussion). *Journal of Bone and Joint Surgery*, **36–B**, 142.

TOUMEY, J. W., POPPEN, J. L., and HURLEY, M. T. (1950): Cauda Equina Tumors as a Cause of the Low-back Syndrome. *Journal of Bone and Joint Surgery*, **32–A**, 249.

ASSESSMENT OF VERTEBRAL OSTEOPOROSIS BY RADIOGRAPHIC AND CHEMICAL METHODS POST-MORTEM

R. A. CALDWELL and D. H. COLLINS, SHEFFIELD, ENGLAND

From the Department of Pathology, University of Sheffield

The measurement of changes brought about by disease is an important step in understanding it, enabling us to determine its incidence and severity under various conditions. A quantitative measurement of osteoporosis is however not easy to obtain. In life a useful histological assessment can be made with care from bone biopsy (Beck and Nordin 1960), but from only a limited field of bone tissue. As ordinarily practised, radiography reveals only advanced degrees of osteoporosis and allows of no accurate comparison of case with case. Barnett and Nordin (1960) have recently recommended indices based upon cortex to shaft ratio in tubular bones and on biconcavity of vertebral bodies. Radiological methods for calculating bone density tend to be elaborate or inaccurate. Although some of the disabilities have been overcome in radiographing peripheral limb bones (Jackson 1951, Virtama 1957; Keane, Spiegler and Davis 1959), the interspersed soft tissue, the position of the bones relative to the tube and the film, and variation in their size are factors that especially prejudice accurate observations on the vertebrae. Even in the post-mortem room, osteoporotic bone atrophy cannot be gauged by the direct measurement of the size and weight of the bones, because the essence of osteoporosis is a rarefaction—that is, a reduction of calcified bone mass in a unit volume of the anatomical bone.

In osteoporosis the trabeculae of cancellous bone become unduly slender and many vanish; those that remain are widely separated. Compact cortical bone undergoes cancellous transformation by the progressive enlargement of resorption cavities which become filled with marrow tissues. The histological picture of osteoporosis indicates an atrophy of the bony framework unaccompanied by osseous regeneration, by fibrous substitution or by notable osteoclastic reaction, as Hadfield observed in reporting on the histological sections in the cases of Burrows and Graham (1945). Osteoporosis has been attributed to the failure of osteoblasts to restore bone lost in the process of physiological resorption (Albright 1947), but Fraser (1959) found no evidence from clinical tests so far available of defective laying down of bone in osteoporosis, and Urist (1958) states that it remains to be proven that the rate of resorption is the same as in normal individuals. Whatever be the case, the bone that remains is calcified bone, but whether individual osteones and trabeculae show abnormally variable degrees of calcification in osteoporosis is a matter yet to be studied by microradiography. Osteoporosis is histologically distinguishable from osteomalacia by the absence of uncalcified osteoid seams with their border of active osteoblasts.

Since we agree with McLean and Urist (1955) that the proper use of the term osteoporosis is to comprehend an increased porosity or " a decrease in the hard portions of bone substance in favour of a relative increase in the soft portions," a measurement of osteoporosis may suitably be expressed in terms of the mass of calcified bone in a certain volume of skeletal structure. For practical purposes this can be reduced to a calculation of the amount of calcium contained in that volume, because the proportion of calcium in bone salt is virtually constant at around 36 per cent (Neuman and Neuman 1958) and it is bone salt that gives strength and rigidity to the skeleton. Before accepting the calcium/volume index as a measure of osteoporosis one must of course be satisfied by histological or other examination that cases of osteomalacia, osteitis fibrosa, Paget's disease of bone, or other bone disease have not been included in the investigation.

Chemical analysis is the direct way of estimating the calcium content of bone. There

are also two indirect methods: 1) the radiographic density, and 2) the specific gravity. In dealing with the vertebrae the radiological method in clinical practice has the inherent difficulties already mentioned and it is, moreover, often impossible to allow for the varying thickness of bone that has been radiographed. In the post-mortem room a uniformly thick slab of bone can be secured and radiographs can be made of it under standard conditions. The specific gravity method was used for some years by one of us (Collins 1959) in the post-mortem room: small cubes of cancellous bone were cut from vertebral centra and dropped into copper sulphate solutions covering a range of specific gravities. The method was considered too crude to continue with, but gave some results in a large series of cases which will be referred to in our discussion.

It is our purpose in the present paper to put forward some results of a study in which the radiographic density and the calcium content of lumbar vertebrae obtained at hospital necropsies have been measured, to indicate the range of calcium contained in these bones, and to show that this can be inferred from the radiograph of the bone slab. Other data

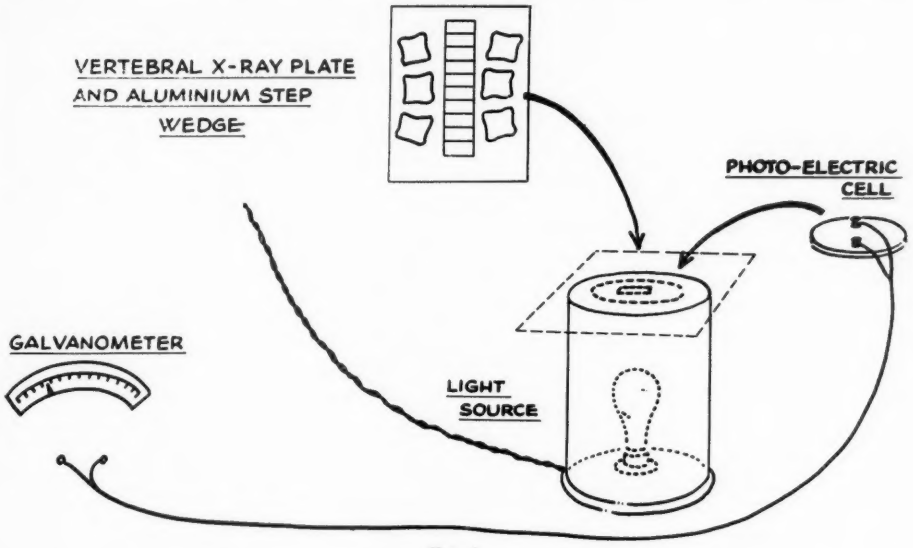


FIG. 1

Diagram of the apparatus used for measuring the radiographic density of specimens of vertebra in terms of thicknesses of the aluminium step-wedge exposed on the same film.

relating to the incidence of osteoporosis among persons dying in hospital and the possible association of other morbid conditions have been gathered, but sufficient material has not yet been examined for a detailed analytical report on these lines.

METHODS

Samples of vertebral bone were obtained in a random series of 100 necropsies on persons dying from various diseases at the Sheffield Royal Infirmary.

A one-centimetre-thick parallel-sided vertical slab of lumbar vertebral bodies and intervening discs, generally including the first lumbar to first sacral vertebrae, was obtained in each case by sawing from the front of the spinal column through to the spinal canal, using two hand tenon saws coupled together with the blades one centimetre apart. The saw cuts were made in such a direction as to include the mid-sagittal plane of the vertebral bodies. After light brushing under running water each specimen was briefly immersed in 1 per cent

gelatin and then suspended in neutral formol-saline. Specimens showing infiltration by tumour or severe Paget's disease were excluded from the series. The bones were kept immersed in fluid until a few moments before radiography. When satisfactory radiographs had been obtained chemical and histological methods were proceeded with.

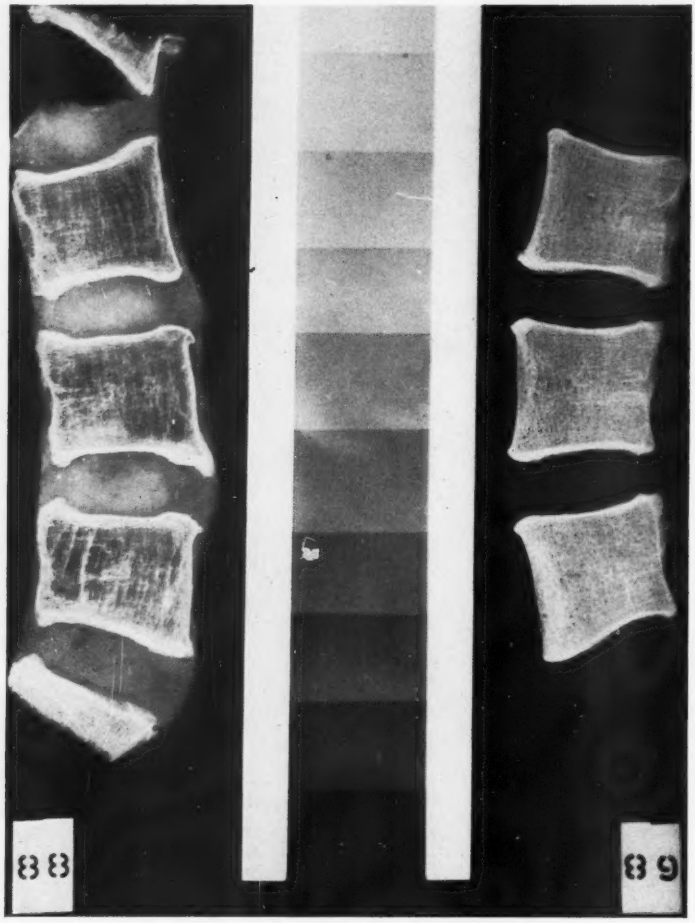


FIG. 2

Reduction of a radiograph of two vertebral slabs and the aluminium step-wedge. The bone slabs are of a uniform thickness of one centimetre. *Specimen* 88 (*on the left*) is from a woman of seventy-two years who had died from acute cholangitis two days after admission to hospital. It shows advanced osteoporosis. The density of the fourth lumbar vertebra corresponds to that of five sheets of aluminium (step-wedge five); the bone contained only 51 milligrams Ca. per cubic centimetre. *Specimen* 89 (*on the right*) is from a man aged forty-three who had died from cerebral tumour five days after admission. The bones being of high normal density contrast well with the other specimen. The step-wedge equivalent of the fourth lumbar vertebra is ten (top step of the wedge); the bone contained 92 milligrams Ca. per cubic centimetre.

Radiography—Wet bone slabs were radiographed alongside an aluminium step-wedge. This was built of one to ten laminae of aluminium sheet 0·3 millimetre thick stepped in such a way as to give a thickness range of 0·3 to 3·0 millimetres numbered from one to ten step-wedge units (Figs. 1 to 3). Constant exposures of ten seconds at 10 kilovolts and 10 milliamps

were made at a tube distance of 63·5 centimetres. Radiographic films (Ilfex $6\frac{1}{2} \times 8\frac{1}{2}$ inches) were exposed in plastic cassettes and developed for five minutes without agitation at 20 degrees Centigrade (Kodak 19B developer).

The density of the radiographic image of each fourth lumbar vertebra (one centimetre

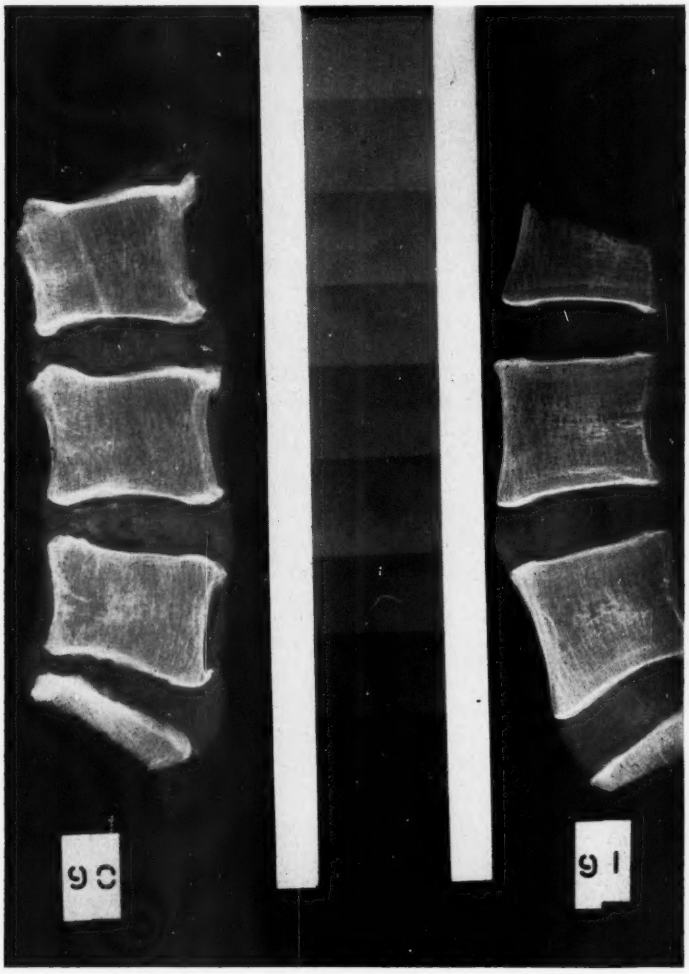


FIG. 3

As Figure 2 but showing vertebrae of average normal density. *Specimen* 90 (*on the left*) is from a woman of fifty-three years who had died from lymphatic leukaemia thirty days after admission. Step-wedge equivalent of the fourth lumbar vertebra is seven. The bone contained 71 milligrams Ca. per cubic centimetre. *Specimen* 91 (*on the right*) is from a man aged sixty-seven who had died within two days from perforated gastric ulcer. Step-wedge equivalent of the fourth lumbar vertebra is also seven, and the bone contained 68 milligrams Ca. per cubic centimetre.

thick) was estimated photometrically in terms of step-wedge units as shown in Figure 1.

Light from an electric lamp (Philips Photocrescenta, 240 volts, 75 watts) was directed through a 2×1 centimetre slot of the image of the vertebral body and, on adjusting the film, through each step image of the step-wedge.

The intensity of the light transmitted in each position was measured by a 6-centimetre photo-cell (E.E.L.) connected to a galvanometer (Pye Scalamp). When the vertebral bodies showed patchy variation of density the average of several readings was recorded; care was taken to observe that each step of the wedge gave a proportionate galvanometer deflection, thus ensuring that each film had been exposed and processed correctly within its sensitivity (Hurter-Driffield) range.

Calcium analysis and calcium/volume index—In addition to the total calcium contained in the bone blocks their wet weight and ashed weight were recorded, but the calcium/ash ratio was found to be more or less constant (*circa* 38 per cent) in all circumstances and the calcium/wet weight ratio does not comprehend the idea of osteoporosis being a reduction of bone mass in anatomical bone structure. For this it was necessary to measure the volume of the intact bone before ashing; this was done by displacement. After radiography, the slab of the fourth vertebral body was dissected from its neighbours and, with a scalpel, stripped of adherent cartilage, blood vessels and connective tissue. Marrow tissues still protected by a film of gelatin remained undisturbed. The block of vertebra was then blotted to remove surface moisture and suspended on one arm of a balance by a fine nylon thread, where it was quickly weighed in air and then in tap water at 20 degrees Centigrade. The difference in the two weights in grammes recorded the volume of the specimen in cubic centimetres. The wet weight of the bone samples ranged from 6·93 to 16·21 grammes and their volume from 5·78 to 13·96 cubic centimetres.

The block of bone, after measurement of its volume, was then ashed in a silica crucible in a muffle furnace at 600 degrees Centigrade for twenty-four hours. The calcium content of the ash was estimated by standard chemical methods. The titration method of Vogel (1951) was used in most cases but later results were obtained with a flame photometer, at first in parallel with the chemical estimations, and then alone.

The assay is expressed as milligrams of calcium per cubic centimetre of bone.

RESULTS

The data in 100 cases are presented here. There were fifty-eight men and forty-two women whose ages ranged from eleven to eighty-eight years. Quantitative results are summarised in Tables I and II and in Figures 4 to 7. Data relating to individual cases are not given, but these were, of course, available for the statistical analyses to which we refer.

The average calcium content of the fourth lumbar vertebral body in the whole series was 68 milligrams Ca. per cubic centimetre volume of bone (range 38–102 milligrams per cubic centimetre). In 75 per cent of cases calcium values fell within the narrower range of 50–84 milligrams per cubic centimetre. Higher calcium contents were observed in younger persons. Thus, the two most heavily calcified vertebrae, with 102 and 101 milligrams Ca. per cubic centimetre respectively, were from men aged thirty-three and twenty-five years and one with 99 milligrams Ca. from a woman aged twenty-nine years.

Men tended to have more calcium in a unit volume of vertebral bone than women. The average in fifty-eight male cases was 72 milligrams Ca. per cubic centimetre (range 47–102) and in forty-two female cases the average was 64 milligrams Ca. per cubic centimetre (range 38–99). Even when allowance is made for the greater proportion of young men than of young women, as in any general hospital necropsy series, the lower calcium content of female bones is still evident at all ages (Table I).

Table I also indicates the decline of calcium in the vertebrae with increasing age, when the figures are expressed as averages. But a wide range of calcium values was still encountered in the older subjects, and with the number of results available it was not possible to calculate a statistically significant regression of calcium on age. A better correlation was achieved between age and radiographic density measured on a ten-degree scale.

Table II shows the average, and range of, calcium content of vertebrae manifesting

radiographic densities of four to ten step-wedge units. No bone was of less than four units density and none greater than ten. A relationship between calcium quantities and radiographic densities is apparent, as may be expected *a priori*, but we were concerned with the possibility of predicting the calcium content from the step-wedge reading, and of seeing whether in doing so it was necessary to take into account the factors of sex, age and number of days in bed, for which information was in each case available.

TABLE I

CALCIUM CONTENT (MILLIGRAMS PER CUBIC CENTIMETRE) OF MALE AND FEMALE LUMBAR VERTEBRAL BODIES EXPRESSED AS AVERAGE MILLIGRAMS PER CUBIC CENTIMETRE IN EACH AGE GROUP

(Number of cases in each group in italics)

	Age group (*years*)					
	10–25	26–55	56–65	66–75	76–85	86–95
Male	82 (*8*)	78 (*13*)	69 (*18*)	66 (*15*)	62 (*4*)	— (*0*)
Female	— (*0*)	70 (*9*)	44 (*12*)	63 (*14*)	59 (*6*)	48 (*1*)

TABLE II

CALCIUM CONTENT (MILLIGRAMS PER CUBIC CENTIMETRE) OF LUMBAR VERTEBRAL BODIES GROUPED ACCORDING TO THEIR RADIOGRAPHIC DENSITY IN STEP-WEDGE UNITS

Radiographic density (*step-wedge units*)	Calcium (*milligrams per cubic centimetre*)		Total number of cases	Male		Female	
	Average	Range		Number of cases	Average age (*years*)	Number of cases	Average age (*years*)
4	48	—	1	0	—	1	88
5	53	38–67	18	8	70·1	10	66·9
6	62	47–76	32	19	60·5	13	68·8
7	75	60–94	31	18	53	13	58·2
8	84	69–101	12	9	37	3	52·3
9	89	71–102	4	2	54·5	2	35·5
10	95	92–97	2	2	29·5	0	—
			100	58		42	

Statistical analysis—Miss Hilda M. Davies, of the Department of Statistics, kindly undertook the analyses and has reported to us as follows.

Multiple regression analyses of calcium on step-wedge reading, age and number of days in bed were carried out for males and females separately, and standard errors of regression coefficients were calculated. In both cases the regression of calcium on age and on days in bed proved to be non-significant: that of calcium on step-wedge reading was very highly significant, and therefore it can be assumed that some estimate of the amount of calcium can be obtained from the step-wedge reading and that in making this estimate there is no need to take either age or number of days in bed into account. This hypothesis was further supported when product-moment correlation and partial correlation coefficients were computed and it was found that the highly significant correlation between calcium and step-wedge reading of about 0·8, both for males and for females, was very little altered by correcting for either the age factor or the days in bed.

To investigate the possibility of a sex difference in such a prediction, a multivariate test of significance (Bartlett 1947) was applied to test the difference between the bivariate mean values of

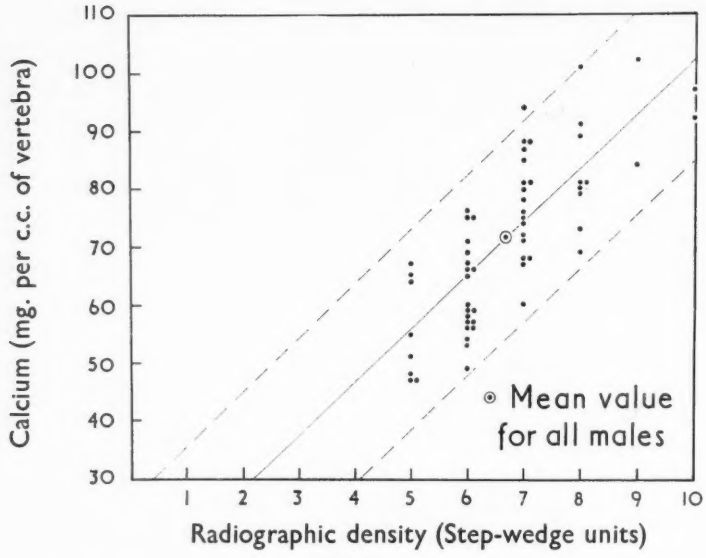


FIG. 4

Regression of calcium on step-wedge (males). Calcium (milligrams per cubic centimetre) plotted against radiographic density (step-wedge units) of lumbar vertebrae, showing regression curve and 95 per cent confidence limits (fifty-eight male cases).

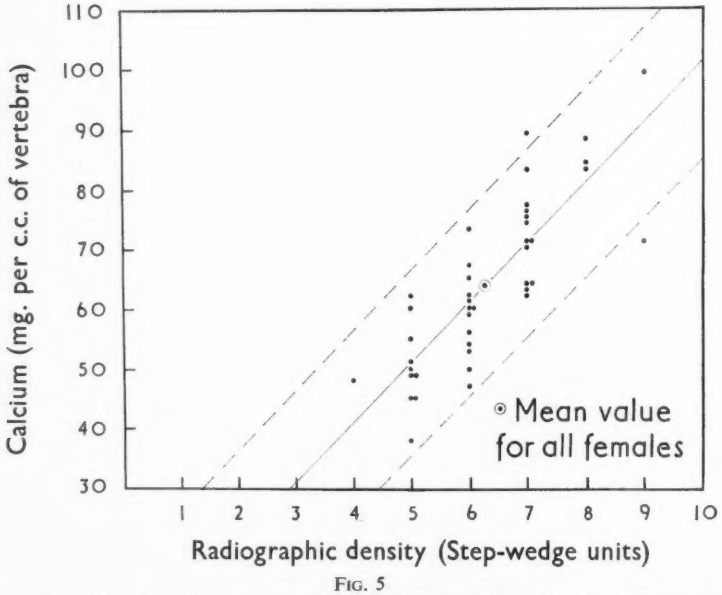


FIG. 5

Regression of calcium on step-wedge (forty-two female cases). As Figure 4.

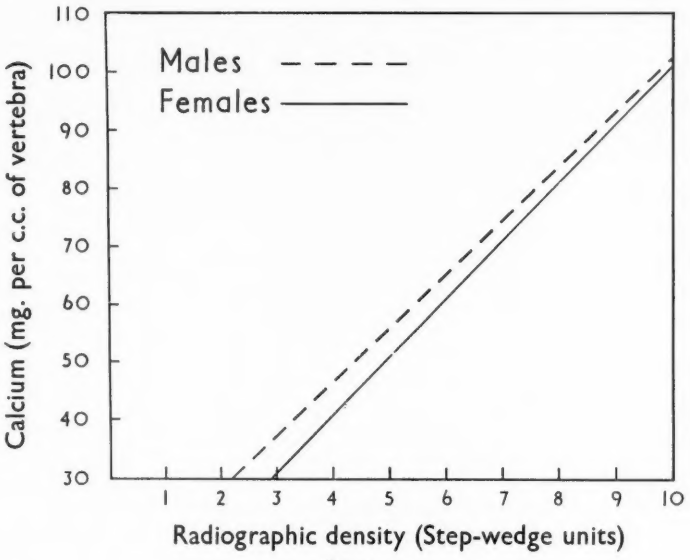


FIG. 6
Regression of calcium content on step-wedge units of radiographic density (males and females).

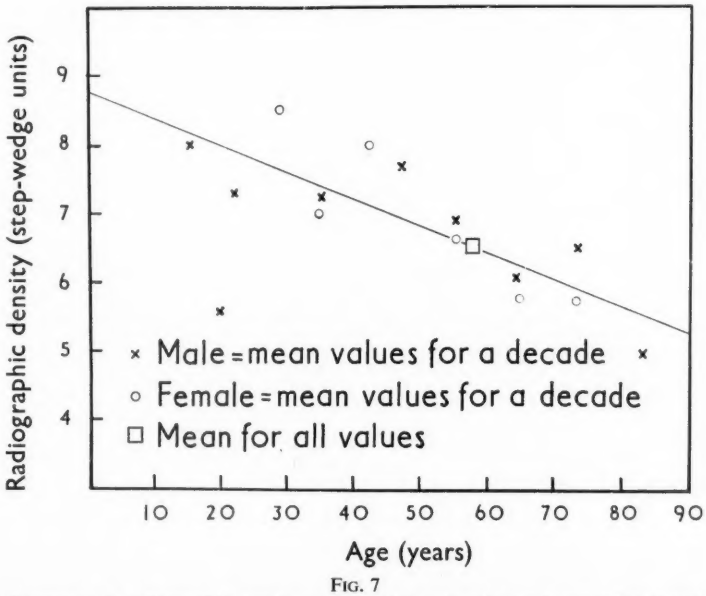


FIG. 7
Regression of radiographic density on step-wedge units with age in years (males and females).

step-wedge reading and calcium for males and females. This gave an F criterion which was significant at the 5 per cent level, suggesting that there is a sex difference. An analysis of variance on calcium and step-wedge data for both sexes combined, testing the effects due to common regression, to difference in location of sex means and to difference in slopes of separate sex regressions, gave no significant difference between the separate slopes but gave an F criterion for difference in location of means which was significant at the 5 per cent level. This bears out the conclusions of the previous test.

Hence it appears that the regressions of calcium on step-wedge reading are parallel for the two sexes but slightly different in position. The separate regression lines of calcium on step-wedge reading for males and females were fitted by the usual method of least squares. The difference between the two regression coefficients was tested against its standard error by the usual Student's t-test and proved not significant, the value of t, in fact, being 0·56 with 96 degrees of freedom, which accorded well with the appropriate value of F in the analysis of variance and bore out the hypothesis that the data are samples from two populations which, though having a different mean value, have the same regression slope.

It seems, therefore, that the step-wedge reading can be used to give a prediction of the amount of calcium present provided that the sex is taken into account and the appropriate regression used. The 95 per cent confidence limits for such a prediction have been calculated in each case.

Figure 4 (males) and Figure 5 (females) show the distribution of calcium quantities in each step-wedge group, the regression of calcium on step-wedge readings and the calculated 95 per cent confidence limits for predicting calcium content from radiographic density in the step-wedge scale here used.

TABLE III

AVERAGE RADIOGRAPHIC DENSITY OF LUMBAR VERTEBRAE OF MALE AND FEMALE SUBJECTS ARRANGED IN TEN-YEAR AGE GROUPS

Age	Male		Female	
	Number of cases	Average step-wedge reading	Number of cases	Average step-wedge reading
0–10	—	—	—	—
11–20	5	8	—	—
21–30	3	7·3	2	8·5
31–40	4	7·25	2	7·0
41–50	6	7·5	2	8·0
51–60	10	6·9	8	6·6
61–70	20	6·2	14	5·8
71–80	8	6·5	12	5·75
81–90	2	5	2	5

Figure 6 compares the regression of calcium on density readings in the two sexes. The statistical analysis revealed that though the difference in slope of the two curves was not significant, the difference in the location of the means was. The implication of this result is not clear; the matter will be referred to in the discussion.

Table III shows the average step-wedge readings of vertebrae in ten-year age groups of men and women, and Figure 7 the regression of radiographic density on age. Miss Davies's statistical report was as follows.

It was noted that the negative product-moment correlations between step-wedge and age were significant and not much influenced by the presence or absence of the other effects, while that of calcium on age with step-wedge held constant was in both cases very small and well below the borderline of significance, implying some relationship between step-wedge reading and age but not

between calcium and age. In view of this an analysis of variance of similar type as before was carried out for the regression of step-wedge on age. This gave a very highly significant value for the combined regression ($F_{1, 96}=45{\cdot}1$) while there was no significant difference for the different sexes between either the separate slopes or the separate locations (F slightly <1 in both cases), implying that there is no sex difference in the effect of age on step-wedge reading but that there is a marked overall regression effect.

The technique of radiographing parallel-sided sagittal slices of the vertebral bodies of uniform (1 centimetre) thickness laid flat on the film cassette enabled us to obtain very clear pictures of the trabecular structure, cortical thickness and contour of the bones. These features are well shown in Figures 2 and 3. In particular the highly rarefied bones of Case 88 in Figure 2 show how in osteoporosis the strong system of little bony plates of the normal vertebral spongiosa is reduced to a delicate web of thin struts, as Collins (1959) demonstrated in macerated specimens. Cortical atrophy is not an important feature of vertebral osteoporosis; the circumferential cortex of a vertebral body is thin in any case, and may become more prominent in osteoporosis—so-called stencilling of outline, by virtue of the greater translucency of the centrum. The bones illustrated in Figure 3 show an average radiographic density and texture. The radiograph of the bone slab gives much more information about cancellous texture than does the thin histological section (Figs. 8 and 9), where many trabeculae are incomplete and cut across obliquely, but such sections have been prepared in all cases in order to exclude osteomalacia, tumour infiltration or other causes of bone rarefaction or sclerosis.

Taking into account a qualitative assessment of the radiological and histological features of each vertebra we conclude that any vertebral slab having a step-wedge reading of five or less with this method shows unequivocal osteoporosis. It will be seen, therefore, from Table I that the incidence of osteoporosis of this degree in this series was 19 per cent. The youngest female to display spinal osteoporosis was aged fifty-one years and the youngest male fifty-nine years.

DISCUSSION

The methods that we have used in this investigation have been appropriate to the study of post-mortem material. They are less relevant to clinical studies, except in confirming that radiographic density of cancellous bone is statistically related to its calcium content (Table II and Fig. 4), a fact that was implicit in Virtama's (1957) radiological investigation of the phalanges post-mortem and was to be expected on *a priori* grounds. By using parallel-sided bone slabs of standard thickness, devoid of superimposed soft tissues, the radiographic measurement by means of a step-wedge and a photo-electric cell becomes much simplified. To counteract the variables of soft-tissue shadows and distance between bone and film in clinical radiography, complex methods must be used, such as those devised by Mack, Brown and Trapp (1949). For our present purposes we are satisfied that the simple radiographic procedures, as outlined in the description of methods and in Figure 1, give a sufficiently accurate prediction of the mineral content of the bone specimens to be used as a method in surveying post-mortem material that is much quicker than chemical analysis. In individual cases some discrepancies between radiological and chemical measurements were encountered (Figs. 5 and 6), and the peculiar fact emerged in statistical analysis that, though falling into the same step-wedge group of density, female bones tended to have slightly lesser amounts of calcium than those of males (Fig. 6). The reason for these discrepancies probably lies in the fact that in a specimen of cancellous bone we are radiographing a system of calcified trabeculae dispersed to a varying degree, depending on the porosity or mesh of the bony network, in soft marrow tissues that also absorb some rays. Mack, Brown and Trapp (1949) stated as a generalisation that calcium is responsible for about 80 per cent of x-ray absorption by bone, but this proportion obviously varies with the ratio of calcified structure to soft-tissue mass.

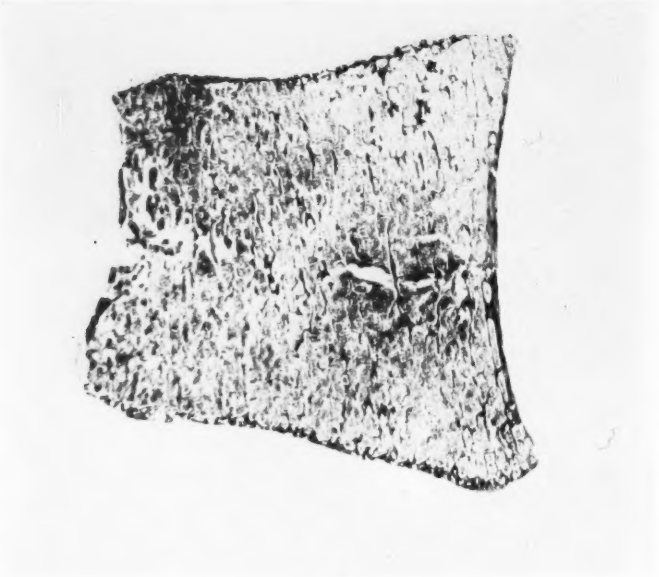


FIG. 8

Histological section of lumbar vertebral body (celloidin-paraffin embedded, stained with haematoxylin and eosin). This bone, of low normal density, showed a radiographic reading of six step-wedge units and a calcium content of 72 milligrams per cubic centimetre.

FIG. 9

Histological section, as Figure 8, of vertebra of high normal density showing a radiographic reading of nine step-wedge units and a calcium content of 90 milligrams per cubic centimetre.

The radiographic scale based on a ten-step aluminium wedge gave in effect only seven degrees of bone density, since no bone was encountered so rarefied as to match with the first three steps of the wedge. Ninety-three of the 100 cases showed densities corresponding with the four step-wedge units five to eight (Table II). The six bones of greater density showed no pathological variation of either radiographic texture (Fig. 2) or of histological structure. The solitary example of a bone showing a step-wedge reading of only four was from an old lady of eighty-eight who on account of pemphigoid had been treated for several weeks before her death with ACTH and prednisolone, which may have accentuated the osteoporotic process. In no other case did we learn of diseases or treatments or other circumstances that might have caused or accentuated osteoporosis.

Radiographs of the bone slabs give a very clear picture of the cancellum of the vertebral bones (Figs. 2 and 3). They also show to what a small extent cortical compact bone enters into the specimen taken in this manner. Indeed, both radiographic and chemical results mainly measure variations in the size, number and degree of separation one from another of trabeculae and differences in the degree of calcification of the osseous rami. It is highly probable that the osseous rami do differ in the extent to which they are calcified. Though osteoporotic bone has not been specially investigated, Amprino and Engström (1952) showed by microradiography not only that the osteones of cortical bone vary in their degree of mineralisation, but that more imperfectly calcified structures are present in spongy than in compact bone. These structures with a low content of mineral salts represent bone in the process of being mineralised (Engström 1956). It seems important, therefore, with a view to establishing the direction of metabolic activity of osteoporotic bone, that the vertebral cancellum should sometime be studied either by microradiography or by interference microscopy (Davies and Engström 1954). In every case, as in the present series, ordinary histological preparations must be examined, because this is the only way of excluding gross variations in the calcium/osteoid relationship, such as pertains in osteomalacia, and of recognising hyperparathyroidism or other pathological causes of bone resorption. In eight of the 100 cases the radiograph revealed a patchy unevenness of the shadow of the lumbar vertebral body. This was unrelated to kyphosis, other deformity or apparent disease. It was not explained, but it did not interfere with our obtaining an average photometric reading of density for the whole bone.

Loss of the transverse trabecular markings and accentuation of the vertical markings is a valuable and constant sign of osteoporosis (Fig. 2). This characteristic was found to correspond with a density reading of five step-wedge units or less, and we are confident in recording as osteoporotic the nineteen cases (eight male, eleven female) showing this degree of rarefaction. The other well known sign of osteoporosis in clinical radiology—that of biconcavity of the vertebral body, the so-called fish vertebra—was in our experience of little value in assessing osteoporosis. It was easy to measure the radiographic profile of the vertebral body and to calculate the ratio of least to greatest vertical depth, as Barnett and Nordin (1960) suggested. Vertebrae in six cases showed this ratio to be less than 75 per cent, a considerable degree of biconcavity; in three the vertebrae were judged to be osteoporotic by other standards, but in the other three cases the vertebrae were of normal density and calcium content. As Collins (1949, 1959) has pointed out, biconcavity results from the pressure of the still turgid intervertebral disc, and if osteoporosis develops after the discs have degenerated or collapsed the vertebral bodies will not become so deformed. Biconcavity, in short, is a reflection of mutual stresses between the vertebral body and its adjoining discs; it is not a function of changes in the bone alone. If biconcavity is conclusive evidence of deformation of a softened vertebral body, then our finding of some biconcave vertebrae of normal density and calcium content can only be explained by assuming that these bones have become remineralised after a period of osteoporosis. But this matter needs further investigation.

The chemical results giving the amount in milligrams of calcium per unit volume (1 cubic centimetre) of anatomical bone are of special interest because, so far as we know, calcium assays have not previously been made of human vertebra, and this method of expressing the results has not been generally adopted in other bone analyses, although it seems to us to be the only way in which the results can be expressed in order to measure what we call osteoporosis and to tally with readings of radiographic density. The range of calcium contents encountered was quite surprisingly wide, from as little as 38 to as much as 102 milligrams Ca. per cubic centimetre of bone, but 75 per cent of cases fell within the narrower range of 50–84 milligrams per cubic centimetre. The average for this series was 72 milligrams per cubic centimetre for men and 64 milligrams per cubic centimetre for women. These figures cannot be taken as averages of general application; much depends on the ages of the subjects comprised in any sample, since it is clear that there is a steady decline with age both of calcium content (Table I) and of radiographic density (Table III and Fig. 7). Even in age groups the range of calcium results is still wide, and the occasional old person may be found with vertebrae that are highly calcified though histologically normal. It was not possible for us to establish a level of calcium content below which a vertebra may be held to be osteoporotic. It was more practicable to name a bone as osteoporotic on radiographic density and texture, other causes of rarefaction being excluded by histological examination. Nevertheless, although there is some overlap of individual results, the generally low calcium of the osteoporotic bones of step-wedge values four or five is apparent in Table II. The average calcium for the eight osteoporotic male cases was 56 milligrams per cubic centimetre as compared with an average of 74 for the fifty male cases judged to show no osteoporosis. In the eleven osteoporotic female cases the average calcium was 50 milligrams per cubic centimetre as compared with an average of 69 for the remaining thirty-one female cases judged to show no osteoporosis.

When the results of calcium analyses of our series of vertebrae are plotted the curve is a smooth one for either sex, though the peak for females is to the left of the male peak (Fig. 10). The skewness of the curves may well be due to the inclusion in our series of a majority of older subjects who tend to show lower calcium values. The main lesson to be learnt from a study of these curves, and of the distribution of the density readings shown in Table II, is that there is no separate peak which might indicate that the sample contained two populations of " normal " and " pathological " bones.

The results indicate the numerical range of mineral disturbances to which the vertebrae are subject. In round figures we may state that the lumbar vertebrae among a mainly elderly hospital population are calcified to the average extent of 70 milligrams Ca. per cubic centimetre, with extreme divergences of 30 milligrams on either side, bones containing 40 milligrams Ca. per cubic centimetre being severely osteoporotic, and bones containing 100 milligrams Ca. per cubic centimetre being mainly among the strong bones of young men. Statements have often been made that a bone must lose at least half its calcified bone mass before it can be recognised as osteoporotic in the clinical radiograph (Fraser 1959). Fusi (1953), who largely excluded the soft-tissue interference by using vertebral body slabs of similar thickness to our own, reported evidence of radiological demineralisation when only 3 per cent of the original calcium content was extracted, and with an extraction of 30 per cent radiological changes were strongly evident; but when the body soft tissues were interposed the extraction of 60 per cent of the calcium was necessary in order to show a notable abnormality. Our analyses partly confirm these findings since, on the average, vertebrae judged to be osteoporotic contained 24 per cent less calcium in the male and 28 per cent less in the female than vertebrae held to be normal. There is, of course, no way of estimating the loss of calcium in any individual bone affected by osteoporosis, because its original calcium content remains unknown. Indeed it is likely that the vertebrae in any individual are most heavily calcified in youth and then proceed to lose calcium (and bone substance) progressively throughout life. It seems that, in osteoporosis, we may be confronted once again, as in osteoarthritis (Collins 1949) and

perhaps, as in hypertension (Pickering 1955), with the problem of determining how and in what manner changes that are the almost universal accompaniment of age are to be distinguished as pathological.

Our present study adds little to our knowledge of the incidence and etiology of osteoporosis. All studies of incidence are based on the somewhat arbitrary and self-imposed standards of the investigators. By our own standards we have called osteoporotic the lumbar vertebrae of eight out of fifty-eight males (14 per cent) and eleven out of forty-two females (26 per cent) in a random series of hospital necropsies. Using entirely different standards, though equally arbitrary—namely, the flotation of cubes cut from the vertebral centra in copper sulphate solution of specific gravity 1·050—Collins (1959) studied a similar but larger series of hospital necropsies and calculated the incidence of spinal osteoporosis as 8 per cent among 189 men and 18 per cent among 147 women aged forty years or more. As in our present series, only one case judged to be porotic was encountered below the age of sixty years.

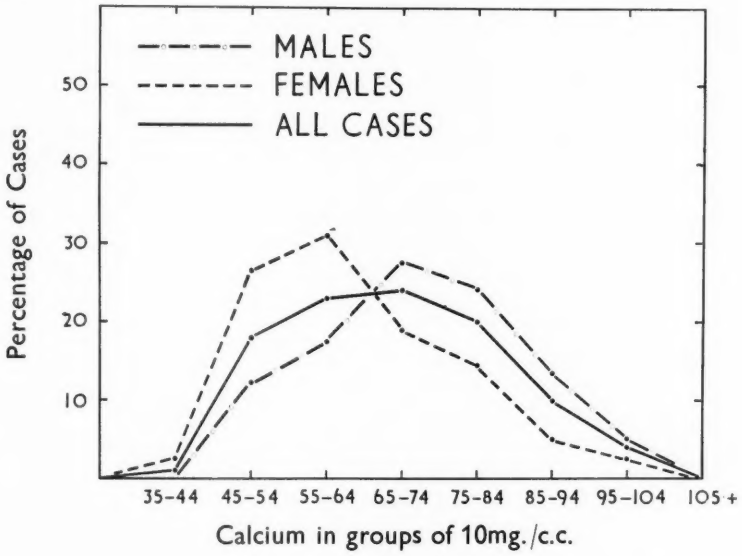


FIG. 10

The percentage incidence of results of calcium analysis of lumbar vertebrae in groups of 10 milligrams per cubic centimetre. Male, female and total cases.

Cooke (1955) reported that osteoporosis is six times commoner in women than in men, but Jackson (1958) found twenty-seven male to eleven female patients with osteoporosis of unknown cause developing in men before the age of fifty-five and in women before the menopause. We have no information of the age at which porosis first developed in our cases but only one patient, a woman aged fifty-one, was younger than sixty-one years at the time of death. The period of confinement to bed during the last illness was noted, but could not be correlated with the state of the vertebrae. The nature of the fatal illness was too varied in this small series of cases to have any apparent bearing on the condition of the bones.

SUMMARY AND CONCLUSIONS

1. Radiological, chemical and histological examinations have been made of the lumbar vertebral bodies in 100 necropsies on patients dying in a general hospital, with a view to determining the range of variation of calcium content and radiographic density in normal and osteoporotic bone.

2. Radiographs were made of sagittal mid-line vertebral body slabs uniformly one centimetre in thickness, and the radiographic density of these specimens was measured in relation to an aluminium step-wedge of one to ten units. Radio-opacity of different vertebrae ranged from four to ten units. The specimen radiographs also clearly revealed the trabecular structure and the lateral profile of the bones.

3. Calcium was chemically estimated and expressed as weight of the element per unit volume of the whole bone mass (that is, of anatomical bone including soft marrow tissue). It ranged from 38 to 102 milligrams per cubic centimetre of bone. In 75 per cent of the cases the range was 50–84 milligrams per cubic centimetre. High calcium values were mostly encountered in young adults, and the calcium per unit volume tended to diminish with age; but a wide range of calcium was still encountered in the older subjects and a better correlation with age was achieved by radiographic density. Both calcium content and radiographic density tended to be higher in the male than in the female bones at all ages.

4. The results of both calcium and radiographic density showed a smooth distribution curve, though skewed through the inclusion in the series of more older people with less mineralised bones; the absence of a double peak in these curves suggests that the examinations were made on a homogeneous population and does not indicate a separate pathological group of osteoporotic subjects.

5. Arbitrary standards must be used to distinguish osteoporotic from normal bones, since neither radiological measurement or chemical assay, nor histological assessment, reveals a point at which the two groups can be separated. In the present series it seemed to us satisfactory to regard as abnormal all bones showing a radiographic density of five or less step-wedge units, and by this standard nineteen of the 100 cases (eight male, eleven female) were deemed to be osteoporotic. Histological examination excluded other forms of bone rarefaction.

6. The regression of calcium on the density measurements proved to be statistically significant and was not affected either by age or by the number of days in bed during the last illness. A small difference between the sexes was apparent, there being slightly less calcium in female than in male bones of equal radiographic density. Provided this is taken into account, the radiographic density scale can be used to predict the calcium content of vertebral bone specimens and should prove a rapid and accurate method in a survey of osteoporosis in post-mortem room material.

We are very grateful to Miss H. M. Davies for the statistical analyses and to Miss D. Kirk, also of the Department of Statistics, for assistance in the calculations. We have received much help from Mr G. Colgrave in the collection of the material and from Mr T. L. Platts in the radiography and photography. Mr A. P. Foster kindly prepared Figure 1.

REFERENCES

ALBRIGHT, F. (1947): The Effect of Hormones on Osteogenesis in Man. *Recent Progress in Hormone Research*, **1**, 293.

AMPRINO, R., and ENGSTRÖM, A. (1952): Studies on X Ray Absorption and Diffraction of Bone Tissue. *Acta Anatomica*, **15**, 1.

BARNETT, E., and NORDIN, B. E. C. (1960): The Radiological Diagnosis of Osteoporosis: A New Approach. *Clinical Radiology*, **11**, 166.

BARTLETT, M. S. (1947): Multivariate Analysis. *Journal of the Royal Statistical Society*, Supplement 9, 176.

BECK, J. S., and NORDIN, B. E. C. (1960): Histological Assessment of Osteoporosis by Iliac Crest Biopsy. *Journal of Pathology and Bacteriology*, **80**, 391.

BURROWS, H. J., and GRAHAM, G. (1945): Spinal Osteoporosis of Unknown Origin. *Quarterly Journal of Medicine*, **N.S. 14**, 147.

COLLINS, D. H. (1949): *The Pathology of Spinal and Articular Diseases*. London: Edward Arnold.

COLLINS, D. H. (1959): Degenerative Diseases. Chapter 4, p. 101 in *Modern Trends in Diseases of the Vertebral Column*, edited by R. Nassim and H. J. Burrows. London: Butterworth & Co. (Publishers) Ltd.

COOKE, A. M. (1955): Osteoporosis. *Lancet*, **i**, 877, 929.

DAVIES, H. G., and ENGSTRÖM, A. (1954): Interferometric and X-ray Absorption Studies of Bone Tissue. *Experimental Cell Research*, **7**, 243.

ENGSTRÖM, A. (1956): Structure of Bone from the Anatomical to the Molecular Level. *Ciba Foundation Symposium on Bone Structure and Metabolism*, p. 3. London: J. & A. Churchill Ltd.

FRASER, T. R. (1959): In Discussion at Clinicopathological Conference on a Case of Bronchiectasis with Osteoporosis. *British Medical Journal*, **ii**, 1,468.

FUSI, G. (1953): Considerazioni e rilievi in tema di osteoporosi con particolare riguardo alle alterazioni scheletriche senili (Studio radiologico e biochimico di un gruppo di 20 vecchi). *Radiologia Medica*, **39**, 209.

JACKSON, H. (1951): Problems in the Measurement of Bone Density. *British Journal of Radiology*, **N.S. 24**, 613.

JACKSON, W. P. U. (1958): Osteoporosis of Unknown Cause in Younger People. *Journal of Bone and Joint Surgery*, **40–B**, 420.

KEANE, B. E., SPIEGLER, G., and DAVIS, R. (1959): Quantitative Evaluation of Bone Material by a Radiographic Method. *British Journal of Radiology*, **N.S. 32**, 162.

MCLEAN, F. C., and URIST, M. R. (1955): *Bone: an Introduction to the Physiology of Skeletal Tissue*, p. 152. Chicago: The University of Chicago Press.

MACK, P. B., BROWN, W. N., Jun., and TRAPP, H. D. (1949): The Quantitative Evaluation of Bone Density. *American Journal of Roentgenology*, **61**, 808.

NEUMAN, W. F., and NEUMAN, M. W. (1958): *The Chemical Dynamics of Bone Mineral*. Chicago: University of Chicago Press.

PICKERING, G. W. (1955): *High Blood Pressure*. London: J. & A. Churchill Ltd.

SACKER, L. S., and NORDIN, B. E. C. (1954): A Simple Bone Biopsy Needle. *Lancet*, **i**, 347.

URIST, M. R. (1958): The Problem of Osteoporosis. *Clinical Research*, **6**, 377.

VIRTAMA, P. (1957): Determination of the Mineral Content of Human Finger Bones by Silver Analysis of Roentgenograms. *Acta Anatomica*, Supplementum 29.

VOGEL, A. I. (1951): *A Textbook of Quantitative Inorganic Analysis*. London: Longman's Green & Co. Ltd.

THE MECHANICAL DISTURBANCES RESULTING FROM LUMBAR DISC SPACE NARROWING

L. M. JONCK, PRETORIA, SOUTH AFRICA

Senior Lecturer, Department of Anatomy, Pretoria University

The syndrome of low back pain constitutes one of the most common diagnostic problems with which the orthopaedic surgeon is presented. A possible etiological factor in a large percentage of these cases, especially those associated with disc lesions, is that of disturbance of the normal mechanism of the spine.

The present investigation was carried out on lumbar spines of the Bantu, and from the information gained in this study one was left with the unmistakable impression that a great deal of misunderstanding exists about the specific function of the supporting structures and the role they play in the mechanism of the spine.

In order to discuss the functional disturbances resulting from disc space narrowing, a sound knowledge of the normal is a pre-requisite. In this paper a survey of the anatomy of the intervertebral disc and its supporting structures will therefore be given. This will be followed by a discussion of the functional disturbances associated with disc space narrowing, because these disturbances may ultimately lead to the so-called disc syndrome.

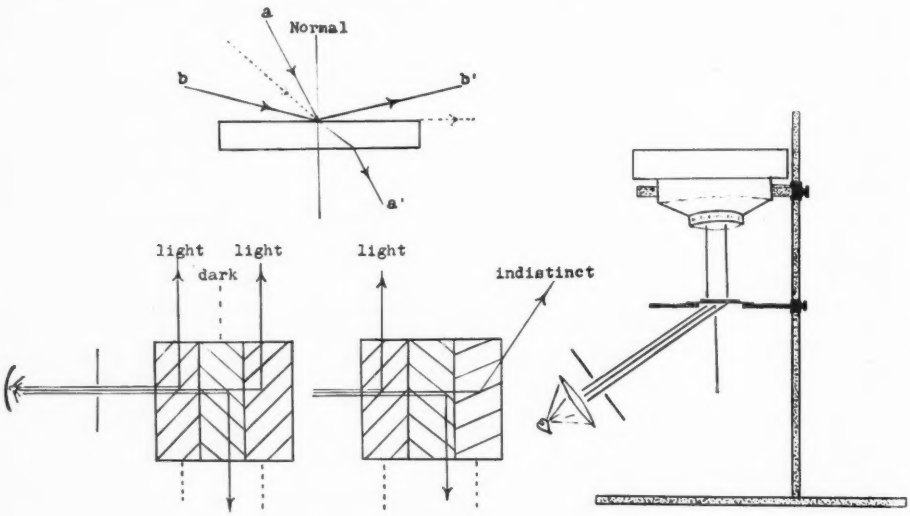


FIG. 1

Diagrammatic representation of light reflection by the fibrous laminae of the intervertebral disc.

MATERIALS AND METHODS

During the past four years 420 Bantu lumbar spines from all age groups were collected from cadavers in the Anatomy Department of the Pretoria University. Thirty of these spines were those of children whose ages ranged from two months to twelve years.

After radiographic examination the ligaments of the vertebral columns were carefully dissected and the relations of the structures forming the intervertebral foramina were noted. Serial sagittal sections an eighth to a quarter of an inch thick were made of a number of these

spines, which proved to be particularly helpful in determining the relations of the nerve root in the intervertebral foramina at different levels.

Frozen sagittal, coronal and horizontal sections (60μ) of the intervertebral discs were made at 1-millimetre intervals. The sections were spread out on glass slides and photographed. They were then studied by reflected parallel light rays.

The reflection of light by the laminae of the annulus fibrosus is a well known phenomenon, and was observed as early as 1835 by Labalt and in 1911 by Fick.

Reflection of light rays by a medium takes place when the angle of incidence is greater than the critical angle (Fig. 1). The fibrous laminae of the annulus fibrosus are light-refractory media. We have developed a technique, based on the principles of light reflection, by which the parallel light rays reflected by the laminae of the intervertebral disc can be photographed. The light source is arranged in such a manner with respect to the section of the intervertebral disc that total reflection takes place. The photographic image of the annulus fibrosus then presents an appearance of alternate light and dark bands. When the light source is gradually rotated the laminae become indistinct; when it is rotated through 180 degrees total reflection again takes place and the previously illuminated layers become dark.

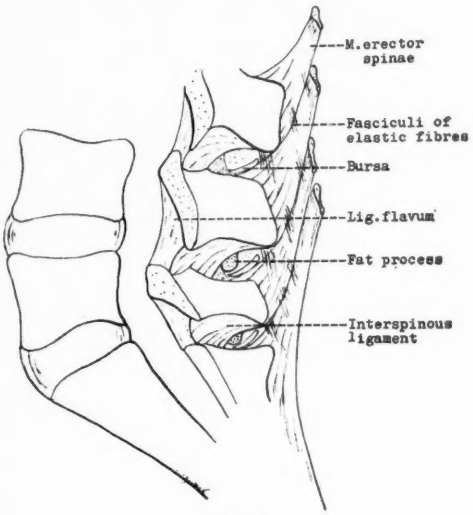


FIG. 2
Diagrammatic representation of the arrangement of the ligaments in the lumbar region of the Bantu.

This technique has many advantages, being relatively simple and requiring no staining, which in itself is a tedious procedure. The photographs are easy to handle, and the method facilitates comparison of the sections of any one disc with sections of discs at other levels.

Total reflection of light by a lamina is possible only when the fibres in a lamina run parallel to each other. Taking into account the sharply defined images that the various laminae produce in the form of white and black bands (Figs. 4 to 9), it becomes obvious that the alternate layers in the annulus run more or less parallel to each other. It also follows that the angle of intersection of two adjacent laminae of the disc is more or less constant.

The structure of the intervertebral disc in some of the related primates was noted; ten monkeys, four baboons and two chimpanzees were examined.

OBSERVATIONS

THE INTERVERTEBRAL DISC AND ITS SUPPORTING LIGAMENTS

The tendinous slips constituting the aponeurotic origin of the erector spinae from the spinous processes of the lumbar vertebrae are held together by fasciculi of short elastic fibres running transversely. These bundles are particularly abundant near the spinous processes (Fig. 2). We were not successful in demonstrating a supraspinous ligament in the lower lumbar region of the Bantu.

The interspinous ligament occupies the interval between two spinous processes. It consists of densely arranged collagenous fibres interspersed with a few elastic fibres. The ligament is attached to the upper borders of the laminae and the base of the spinous process of a lower vertebra, and runs obliquely upwards and backwards to the spinous process of

the adjacent upper vertebra. Anteriorly it blends with the ligamentum flavum. The thickness of the interspinous ligament increases progressively from the lower thoracic region to the lower lumbar region.

The ligamentum flavum occupies the interval between two adjacent laminae. Its thickness likewise increases progressively from above downwards in the vertebral column. Laterally the ligamentum flavum enters into the formation of the anterior part of the capsule of the paravertebral joint and so bounds the intervertebral foramen posteriorly.

The intertransverse ligaments bridge the intervals between the transverse processes of the lumbar vertebrae. Each ligament consists of two parts, lateral and medial. In the intertransverse spaces of the upper lumbar vertebrae the lateral part is poorly developed and hardly justifies description as a ligament. It has the appearance of a thin fascial layer which separates the muscles lying in front of the transverse process from those behind it (Fig. 3). In the lower two intertransverse spaces it contributes to the ilio-lumbar ligament.

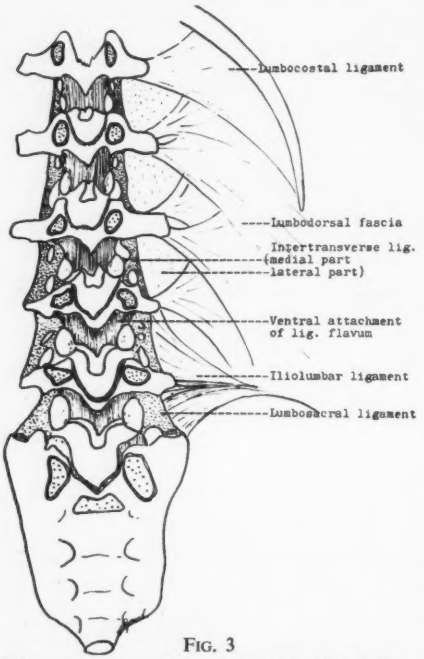


FIG. 3

Diagram to show the attachments of the ligamentum flavum to the ventral aspects of the laminae of the lumbar vertebrae, and the ligaments in the intertransverse spaces. It also clearly demonstrates the relationship of the ilio-lumbar ligament to the lumbo-dorsal fascia.

The medial part of the ligament is arranged in the form of a tendinous arch which provides protection for the posterior primary ramus and blood vessels. The thickness of this medial part increases progressively from above downwards in the vertebral column. The part between the transverse processes of the fifth lumbar vertebra and the sacrum is sometimes described as the lumbo-sacral part of the ilio-lumbar ligament (Fig. 3).

The ilio-lumbar ligaments extend between the transverse processes of the last two lumbar vertebrae on the one hand and the iliac crest and anterior aspect of the upper part of the sacrum on the other. The fibres of the ilio-lumbar ligaments radiate from the transverse processes of the fourth and fifth lumbar vertebrae, and correspond to those of the deep layer of the lumbo-dorsal fascia. The anterior fibres pass obliquely downwards and are attached to the posterior half of the inner lip of the iliac crest. Occasionally they form a hard sickle-shaped bundle with the concavity anteriorly.

The anterior and posterior ligaments are broader and stronger in the thoracic than in the lumbar region. These ligaments are not attached to the intervertebral discs, but gain attachment to the vertebral bodies beyond those parts developed from the epiphysial ring (Figs. 8 and 9).

The capsular ligaments consist mainly of collagenous fibres. On the dorsal aspect of the paravertebral joints they are extremely thin. In the lower lumbar region their inferior parts are strengthened and partly replaced by the interspinous ligaments. Anteriorly these joints are covered by the ligamentum flavum.

The important constituent parts of the intervertebral disc are the annulus fibrosus and the nucleus pulposus.

The adult annulus fibrosus is composed of a series of laminae which form incomplete rings. The fibres of each lamina follow an oblique course between two vertebrae and cross the fibres of an adjacent lamina at a certain angle. The angle of intersection varies between

30 and 60 degrees depending on the state of the disc and the pressure to which it is subjected—for example, whether the individual is recumbent or standing erect, and whether the disc is subjected to muscular force or weight bearing. The angle of intersection for any one disc is more or less constant under a given set of circumstances.

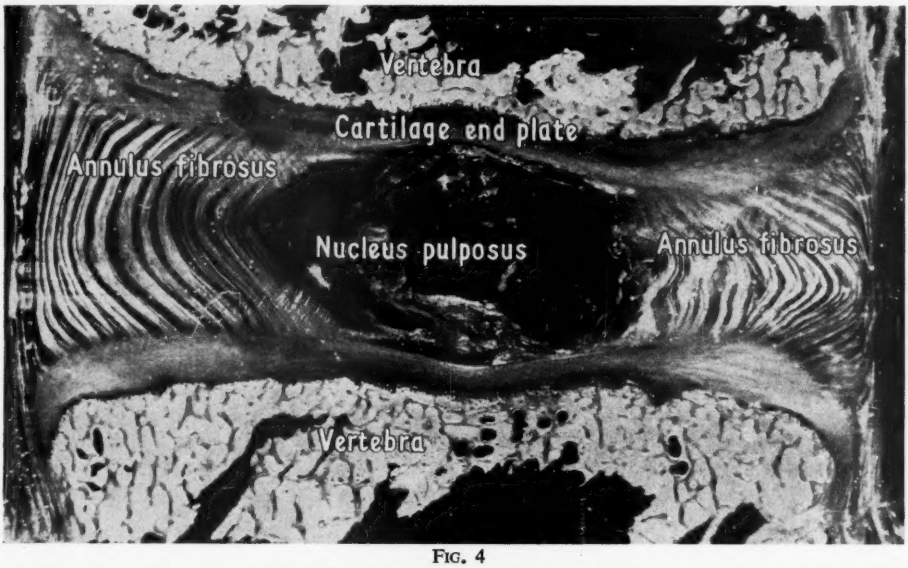


FIG. 4

Sagittal section of intervertebral disc of a child of eight. It demonstrates the gel consistency of the nucleus pulposus. The alternating light and dark bands represent the laminae of the annulus fibrosus. Note the sharp contrast between nucleus pulposus and annulus fibrosus.

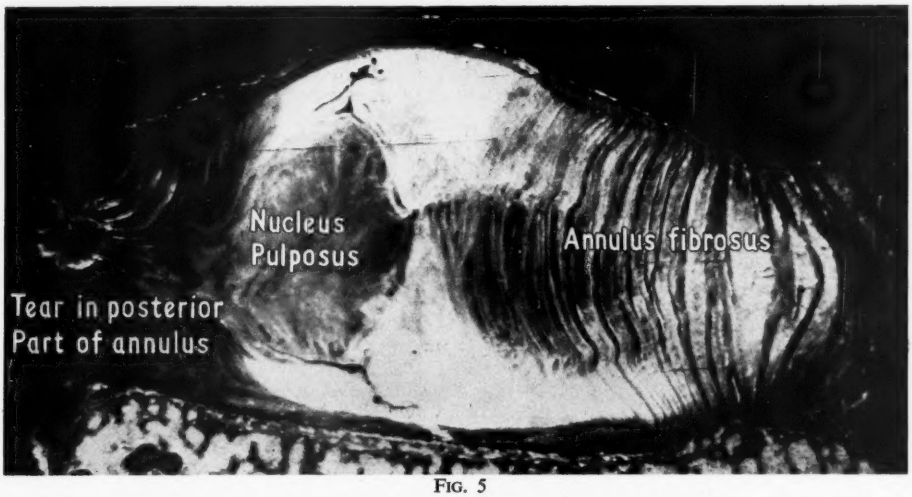


FIG. 5

Sagittal section showing traumatic herniation of the nucleus pulposus in an adult aged thirty. The gel consistency of the nucleus pulposus is replaced by a fibrillar network.

The nucleus pulposus consists of a fibro-gelatinous material. It is confined to its anatomical shape and position by the surrounding annulus fibrosus and the overlying cartilaginous end-plates. It consists of a homogeneous ground substance with a fine fibrillar

structure with no definite arrangement (Fig. 5). In the young child the structure of the nucleus pulposus stands in sharp contrast to that of the annulus fibrosus, due to the gelatinous consistency of the nucleus in this age group. In the higher age groups the difference between

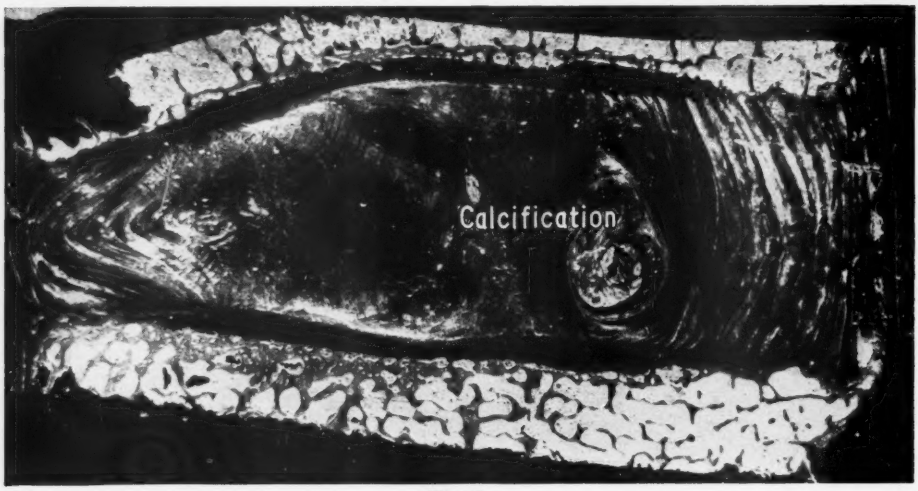


FIG. 6
Calcification in an intervertebral disc of an adult aged forty.

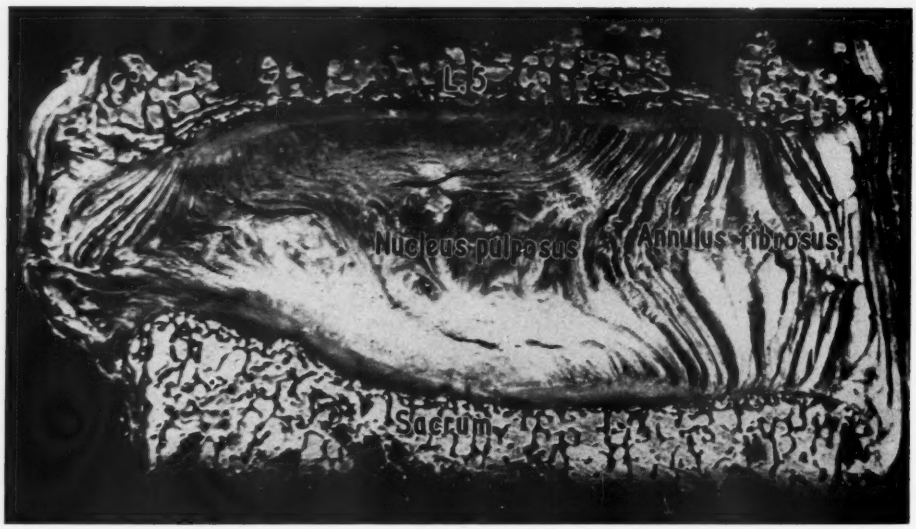


FIG. 7
Sagittal section of the lumbo-sacral disc of an adult aged fifty, demonstrating that the posterior part of the intervertebral disc occupies the lower part of the intervertebral foramen.

the nucleus pulposus and the annulus fibrosus is less conspicuous because the fibres of the nucleus become coarser (Figs. 4 to 8).

At different ages variations exist in the density of the nucleus pulposus. The density of the nucleus was found to increase with age, but we were not successful in establishing norms for various ages.

THE ARTICULAR PROCESSES AND THE INTERVERTEBRAL FORAMINA

The articular processes take part in the formation of the posterior boundary of the intervertebral foramina. The part of the ligamentum flavum substituting the capsular

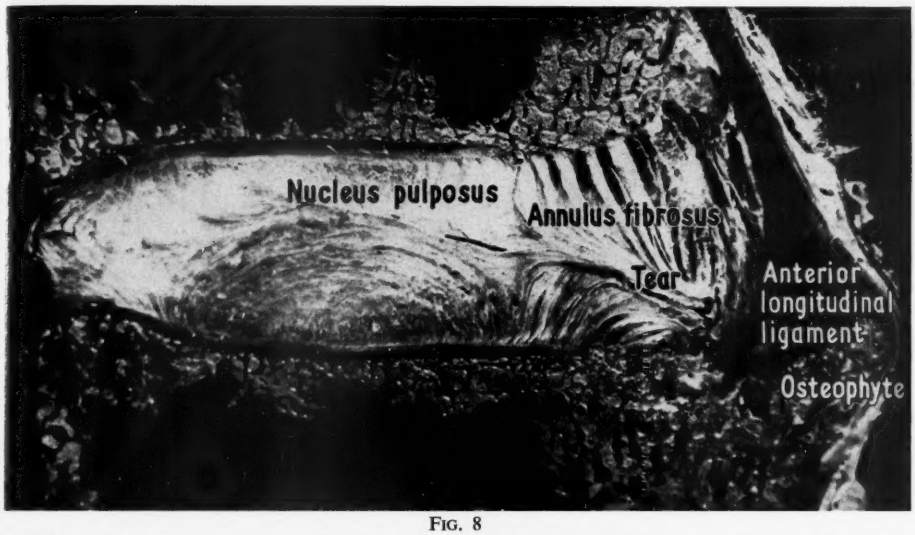


FIG. 8

Sagittal section of the intervertebral disc of an adult aged sixty with a tear in the anterior part of the disc and an osteophyte developing in the anterior longitudinal ligament.

FIG. 9

An osteophyte in the anterior longitudinal ligament of a subject aged seventy. Note the density and the almost homogeneous appearance of the nucleus pulposus with hardly any contrast between nucleus pulposus and annulus.

ligament covers the articular process and is closely related to the nerve root in the intervertebral foramen.

The inclination of the articular processes varies in the different lumbar segments. The processes of the sacrum and the superior articular processes of the fifth lumbar vertebra are inclined forwards, whereas the articular processes of the fourth lumbar vertebra are more or

less vertical. In the upper lumbar region the superior articular processes are inclined backwards (Fig. 10).

The relationship of the posterior surface of the disc and the articular processes also varies in the different lumbar segments. In the lower lumbar region, with the person standing erect, the posterior aspect of the disc, especially that of L.5–S.1 and to a lesser extent that of L.4–5, lies in contact with the anterior aspect of the superior articular processes of the lower vertebra. This part of the intervertebral disc therefore normally occupies the lower part of the intervertebral foramen (Fig. 10).

The relations of the nerve root in the intervertebral foramen are variable and depend on the age of the individual and the segmental level of the foramen. In the embryo the nerve root lies horizontally; in the child it passes obliquely downwards, and the obliquity increases with age.

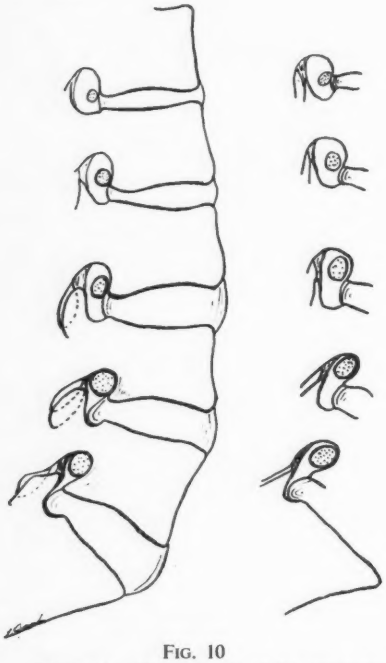


FIG. 10

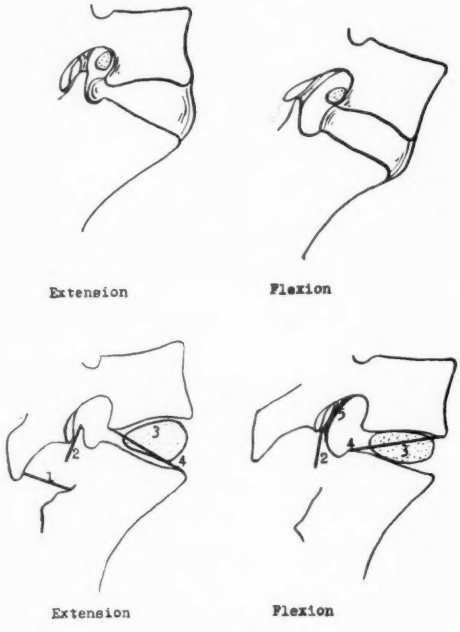


FIG. 11

Figure 10—The relations of the structures forming the intervertebral foramina at the different lumbar levels in erect posture. Figure 11—Analysis of the structures playing a role in guiding and limiting the movements of flexion and extension. 1) Interspinous ligament. 2) Articular surfaces of the paravertebral joints. 3) Nucleus pulposus. 4) Fibrous laminae of the annulus fibrosus. 5) Ligamentum flavum.

The variation in the relationship of the nerve roots in their intervertebral foramina at the different disc levels can be explained on the basis of the angulation of the vertebral bodies to produce the lumbar lordosis. This angulation, which is most accentuated in the lower lumbar region, causes an anterior inclination of the superior articular processes in this region, whereas they are practically vertical at the higher lumbar levels.

In the intervertebral foramina of the lower lumbar region, and especially in the case of L.4–5 and L.5–S.1 levels, the nerve lies immediately above the disc and grooves the postero-lateral aspect of the vertebral body directly below the pedicle of the upper vertebra. This groove is visible on the lateral radiographs of most lumbar spines.

The intervertebral foramina are smaller in the lower lumbar region than in the upper lumbar region. The size of the foramina increases with flexion and diminishes with extension.

THE FUNCTIONS OF THE INTERVERTEBRAL DISC AND ITS SUPPORTING STRUCTURES

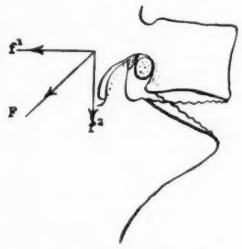
Under normal conditions with an intact nucleus pulposus flexion and extension take place between two vertebrae as a rocking or rotating movement over a ball, the nucleus pulposus. These movements are controlled and guided by the fibrous laminae of the annulus fibrosus, and by the fibres of the interspinous ligament and ligamentum flavum which run in the corresponding directions (Fig. 11). These movements can aptly be described as simulating those of a rocking-horse.

The interspinous ligaments and the fibrous laminae of the annulus fibrosus running in the corresponding direction are not only important factors in determining the course an upper vertebra will follow on a lower one in the extension movement, but the direction in which the fibres run also enables these structures to prevent posterior displacement of the upper one of any two vertebrae in the lumbar region. These ligaments are therefore important in preventing retro-spondylolisthesis. If the ligaments were absent or lax retro-spondylolisthesis would undoubtedly occur in extension in consequence of the strong muscular traction exerted by the erector spinae group.

In flexion the ligamentum flavum becomes tense and forward slipping of an upper vertebra is prevented by it as well as by the fibrous laminae of the annulus fibrosus running in the appropriate direction.

The movement of lateral flexion is in the main checked (guided and limited) by the fibrous laminae in the anterior and posterior parts of the disc. These fibres of the annulus fibrosus are aided by the ilio-lumbar ligament and the strong medial parts of the intertransverse ligaments, especially in the lower lumbar region.

The different fibrous laminae of the annulus fibrosus do not intersect but lie one on the other, and run in different directions. This cruciate arrangement forms an essential part of the shock-absorbing mechanism of the disc. It also confers on the annulus fibrosus the power of controlling torsional movements.



Disc space narrowing without displacement

FIG. 12

Disc space narrowing with displacement

FIG. 13

Figure 12—Diagram representing narrowing of a disc (without displacement) causing laxity of the laminae of the annulus fibrosus. F is a muscular force acting on the fifth lumbar vertebra: it consists of two components, f^2 a vertical component, and f^1 a horizontal component which is mainly responsible for posterior displacement. Figure 13—Diagram representing impingement of the articular process of the sacrum on the pedicle above with consequent bone sclerosis and probable pressure on the nerve root.

In extension, as when the subject stands erect, the intervertebral disc bulges posteriorly and inferiorly and thus cradles itself in the superior notch of the vertebra below; this is most pronounced in the case of the lumbo-sacral disc (Figs. 10 and 11). In this position the intervertebral foramina are smallest and in flexion they are largest. The greatest variations in the size of the intervertebral foramina during these movements take place more especially in those segments which contribute the most to a specific movement of the spine.

DISCUSSION

In most anatomical text-books, including those of Fick (1911), Rauber-Kopsch (1955), Grant and Smith (1953), Braus-Elze (1954), and Gray (1958), the descriptions of the supraspinous ligament are more or less similar. The ligament is said to extend from the spinous process of the seventh cervical vertebra to the medial sacral crest. It is described as being well developed in the lumbar region and as consisting of longitudinal collagenous fibres, of which the deeper fibres connect adjacent spinous processes. The superficial fibres extend over three or four segments.

We were not successful in demonstrating a ligament of this description in the lumbar spine of the Bantu. We have found, in fact, that the aponeurotic origin of the erector spinae from the spinous processes in the lumbar region could easily be mistaken for a supraspinous ligament.

The tendinous slips constituting the aponeurotic origin of the erector spinae lie close to each other and are held together by transversely running bundles of elastic fibres. Those elastic fibres close to the spinous processes are well developed and connect adjacent tendinous processes close to their origin from the spinous processes (Fig. 2). These elastic fibres may simulate a supraspinous ligament. However, they do not connect adjacent spinous processes but rather form a connection of either adjacent tendinous slips or of a tendinous slip and a spinous process.

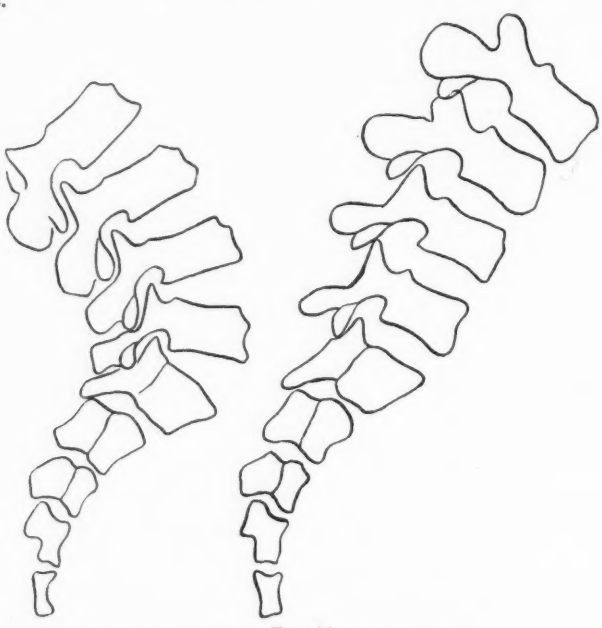


FIG. 14

Tracings of radiographs of flexed and extended spine illustrating widening of the interspinous spaces.

From a functional and mechanical point of view the distance by which the spines of contiguous vertebrae are separated in the movements of flexion and extension does not justify the assumption of the existence of a strong collagenous supraspinous ligament. A ligament of this description would serve only to hamper the movement of flexion or would rupture. A fibrous structure with vertically running fibres cannot be reconciled with the type of movement that takes place between two segments (Fig. 14).

The supraspinous ligament of the thoracic region may be looked upon as the remains of the spinous origin of the erector spinae in this region. This postulation is conceivable when one considers the structure of the spinales column of the erector spinae. It is furthermore a well known fact that a muscle may undergo regressive changes and continue its existence in the form of a ligament (Huntington 1902).

The interspinous ligament was described by Weibrecht (quoted by Fick 1911) as a thin membrane which separates the muscles of the back like an intermuscular septum. Von Luschka (1867) described it as a fibrocartilaginous mass comparable to the annulus fibrosus. The description of the interspinous ligament by Braus-Elze (1954) and Gray (1958) is in accordance

with the description by Fick and Bardeleben (1904) and is represented diagrammatically in Figure 17. According to Rauber-Kopsch (1955) the interspinous ligament occupies the interval between two spinous processes and extends from the base of one spinous process to the apex of the next spinous process (Fig. 18). Grant and Smith (1953) described a criss-cross arrangement of the fibres forming the interspinous ligament (Fig. 19).

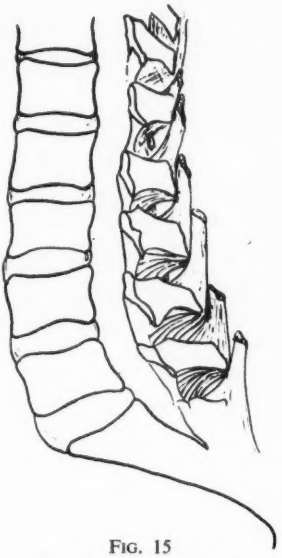


FIG. 15
The South African Bantu.

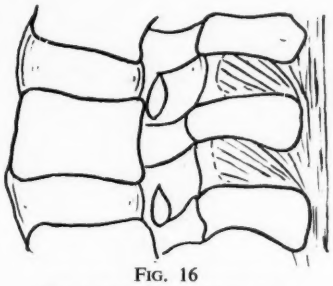


FIG. 16
Der Anatomie des Menschen.
K. v. Bardeleben. (1904–1910.)

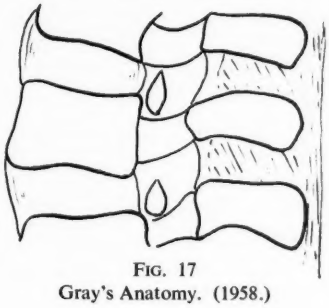


FIG. 17
Gray's Anatomy. (1958.)

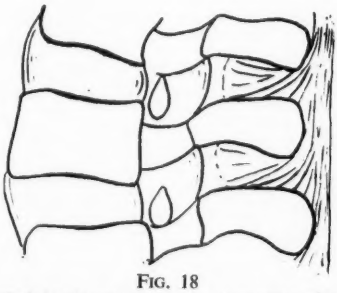


FIG. 18
Lehrbuch und Atlas der Anatomie des Menschen. Rauber-Kopsch. (1955.)

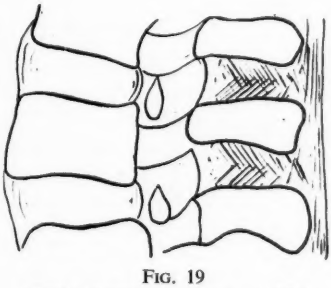


FIG. 19
Morris Human Anatomy. (1953.)

FIGS. 15 TO 19
Diagrammatic representation of the ligaments of the Bantu lumbar spine as compared with classical descriptions.

From the above descriptions it is clear that there is a divergence of opinion on the exact structure of the interspinous ligament. Nevertheless, all agree that the ligament is best developed in the lumbar region.

In the Bantu the interspinous ligament is not limited to the interspinous space, but has an extensive attachment to the whole of the superior margin of the lamina and the base of

the spinous process of a lower vertebra. The direction in which the fibres run enables it to counteract any excessive muscular force which comes into play when the erect attitude is assumed. In the lower lumbar region the interspinous ligament contributes to the stability of the lumbar spine.

The factors that enable the disc to cope with the forces involved in gaining and maintaining the erect posture, as well as the considerable stresses to which it is subjected in the upright posture, are mainly: 1) The arrangement of the fibrous laminae of the annulus fibrosus; 2) an intact nucleus pulposus; and 3) the surrounding tissue elements.

The cruciate arrangement of the fibrous laminae of the annulus fibrosus is not a new discovery; it was described as long ago as 1835 by Labalt, who also noted that the angulation of the fibres varies in different regions of the vertebral column. The findings in the present study agree with those of Labalt. As the earlier observations were, however, given in the form of general statements, it was considered that an understanding of the true mechanism of the intervertebral disc demanded a more detailed examination of its fibrous system.

By means of the reflection of parallel light rays by successive laminae of the annulus fibrosus it was possible to demonstrate that the angle of intersection of the various fibrous systems is more or less constant for any one disc.

This finding is of practical significance in so far as a difference in angulation between the fibrous laminae in the same disc presupposes the presence of shorter fibres which would be subjected to greater strains than the longer fibres. An arrangement of this nature would not be suitable for the type of movement taking place between two vertebrae, and may lead to radial tears, which can in fact at times be demonstrated in an otherwise normal intervertebral disc.

Some authorities (Walmsley 1953) believe that the criss-cross arrangement of the annular laminae is to be primarily correlated with the internal pressure exerted on them by the nucleus pulposus. We are of the opinion that the structure of the annulus fibrosus is closely associated with the functions of the spine, whether it concerns a quadrupedal or a bipedal organism. In our series no difference was demonstrated in the structure or in the construction of the intervertebral discs of quadrupeds as compared to bipeds.

The proper functioning of the nucleus pulposus depends upon the integrity of its limiting structures, namely the cartilage end-plates above and below, and its surrounding annulus fibrosus.

It is generally accepted that the semi-fluid character of the nucleus pulposus allows it to adapt itself readily to changes in its form during movement of the vertebral column and thus to function normally (Walmsley 1953).

Although the disc has a relatively small surface area it is subjected to a variety of stresses, often of considerable magnitude. It has been estimated by Hoad and Rosenberg (1953) that the fourth lumbar intervertebral disc is subjected to a total load of 1,500 lb. when a weight of 100 lb. is lifted with the trunk flexed.

Franceschini (1947) referred to the oblique course of the fibres in the posterior part of the annulus which renders them subject to compression force. According to Walmsley (1953), the nucleus pulposus is responsible for the even distribution of the compression forces over the entire upper and lower surfaces of the vertebral bodies. He further noted that in any particular position of the vertebral column, the compression forces will be greatest on those parts of the vertebral bodies that bound the concave side of the curvature.

Brown, Hansen and Yorra (1957) noted that the greatest amount of bulging occurred in the anterior third of annulus fibrosus, an observation which favours direct compression of the annulus fibrosus rather than bulging caused by deformity of the nucleus pulposus. According to these authors it seems that the disc is subjected primarily to direct compression rather than to hoop tension. They concede that the behaviour of the disc may vary with age. Friberg (1948) concluded that the annulus also had a weight-bearing function.

Happey, Horton, MacRae and Naylor (1955) drew attention to the ability of the nucleus pulposus to convert a vertical pressure into a horizontal thrust on the annulus fibrosus. It has furthermore been suggested that the annulus fibrosus is elastic in order to dissipate the resultant thrust from the nucleus. This elastic mechanism is provided for by the criss-cross arrangement of the annulus fibrosus (Horton 1958).

It is the function of the intervertebral disc to transfer the forces acting on the vertebra above to the one below, and this is efficiently brought about by the nucleus pulposus, a structure which complies with the laws applicable to a fluid under a compressive force. The force to which the nucleus pulposus is subjected is transmitted in all directions and affects the cartilage end-plates as well as the annulus fibrosus, which represent the limiting structures of the nucleus.

If we assume that the load to which a disc is subjected is uniformly distributed over the upper surface of the vertebra below, then the load carried by each of the two elements, the annulus fibrosus and the nucleus pulposus, is proportional to their respective areas of contact on the surface of the vertebra. The area of the central part of the cartilage end-plate which is related to the nucleus pulposus varies between 24 per cent and 33 per cent of the total surface affected by weight transmission. This means that about a quarter or one third of the load is transmitted by the nucleus pulposus as a compression force. The remainder of the load is transmitted by the annulus fibrosus.

Since the nucleus pulposus complies with hydrostatic laws it is incompressible but subject to deformity, which is counteracted by the limiting capsule, the annulus fibrosus. The deformation of the sphere generates hoop tension in the obliquely set fibrous laminae of the annulus fibrosus. The vertical component of this tension force represents the remainder (76 per cent to 66 per cent) of the load.

In view of these features of the nucleus pulposus it appears that the nucleus pulposus provides the mechanism responsible for the maintenance of balance between hoop tension and compression force.

When disc space narrowing occurs, with loss or diminution of the contents of the nucleus pulposus, important changes take place in the capacity of the intervertebral disc to transmit weight. The annulus fibrosus is now no longer subjected to hoop tension, but to a direct compressive force.

A construction such as the annulus fibrosus which is effective in hoop tension is not necessarily effective under a compressive force. This change in the mechanics of weight transmission is an important factor in the causation of further degeneration of the annulus fibrosus with consequent progressive disc narrowing. The cartilage end-plates overlying the nucleus pulposus are comparable to articular cartilage such as is found within the knee and hip joints and which is effective in withstanding compressive forces.

As the intervertebral disc at the L.5–S.1 level degenerates, the fifth lumbar vertebra approximates to the sacrum, with consequent increase in the overlap of the paravertebral articular facets, the superior articular process encroaching upon the pedicle above it (Fig. 13). The part of the ligamentum flavum attached to the front of the inferior articular process is dissected away from its original attachment and now projects as a bulge in front of the superior articular process. In a large percentage of the anatomical specimens in which disc space narrowing was found, the part of the ligamentum flavum related to the paravertebral articulation was found to have completely disappeared, probably due to the excessive lip formation of the articular processes, or due to pressure atrophy.

In disc space narrowing the vertebral body not only descends, but there is also a tendency for posterior displacement of the fifth lumbar vertebra on the sacrum (Fig. 13). Two factors may be responsible for this backward movement of the upper vertebra. First, the superior articular processes in the lower lumbar region are inclined backwards at an angle. Secondly—and this is the more important factor—the powerful muscular force which under normal

conditions would have acted on the fifth lumbar vertebra to stabilise it on the sacrum in the erect posture draws the vertebra backwards. In consequence of the loss of contents of the intervertebral disc, the interspinous ligaments and the fibrous laminae of the annulus fibrosus running in the corresponding direction become lax and are no longer capable of counteracting this muscular force (Fig. 12).

As a result of these changes the movement between the two vertebrae is now no longer a rocking movement following a fixed course as determined by the complex of the interspinous ligament, annulus fibrosus, articular processes, ligamentum flavum and nucleus pulposus. Instead a backward and forward sliding movement occurs which not only increases the destructive effect on the intervertebral disc, but also puts an extra load on the articular processes, with consequent arthritic changes.

These findings support the views of Franceschini (1947), Friberg (1948), Walmsley (1953), and Lindblom (1952) that fibrous tissue and fibrocartilage are pre-eminently adapted to withstand tensile forces, and that degenerative changes may occur within them when they are subjected to compression. This process, which Lindblom (1952) considers a pressure atrophy, is certainly responsible for the progression of disc space narrowing, but in the author's opinion the primary factor which initiates the process is the diminution or loss of contents of the nucleus pulposus; the latter may be the result of trauma, disease, or even the result of the maturation process to which all discs are subjected.

In this investigation well over 400 lumbar spines were subjected to a critical anatomical examination of the intervertebral disc and its relation to the intervertebral foramen. In not one case was there any evidence that the disc had exerted pressure on the nerve root. In those spines with disc lesions it is very probable that direct pressure on the nerve root had been exerted by the part of the ligamentum flavum lying in front of the superior articular process; after the disappearance of this part of the ligamentum flavum the superior articular process may have exerted pressure directly on the nerve.

As the contents of the intervertebral disc diminish, the narrowing of the disc may progress until eventually the cartilage end-plates lie almost in apposition.

In this series the impression was gained that the nerve root accommodates itself in cases of gradual disc narrowing by carving a niche for itself on the postero-lateral aspect of the vertebral body below the pedicle, and thus becomes protected from further pressure. This mechanism is under investigation. Furthermore, because of the greater anterior dimensions of the lumbo-sacral disc, the fifth lumbar vertebra is tilted forwards when narrowing occurs at this level. This forward tilting of the fifth lumbar vertebra on the sacrum will lead to alleviation of the tendency to nerve pressure, and may be an explanation why narrowing of the disc may occur without pressure symptoms.

SUMMARY AND CONCLUSIONS

1. In this investigation one was impressed by the close relationship that exists between the constituent parts of the intervertebral discs and the surrounding supporting structures. The part that is responsible for the maintenance of a co-ordinated balance between these structures, and hence for the effective mechanism of the spine, is the nucleus pulposus.

2. The cruciate arrangement of the annulus fibrosus is related to spinal function, and the angle of intersection of consecutive laminae of the annulus fibrosus is more or less constant.

3. In the Bantu it was found that the interspinous and supraspinous ligaments differ from the classical descriptions given in anatomical text-books.

4. The movements taking place between two vertebrae are comparable to those of a rocking-horse.

5. Narrowing of a disc is a progressive process which is the result of the disturbances in the balance between the components of the force to which a disc is subjected. The process is initiated by the loss of nucleus pulposus content.

6. With the approximation of two adjacent vertebrae a disturbance of the relations of the structures in the intervertebral foramen was noted. On anatomical grounds, when a disc is narrowed it appears that the most likely structures to cause pressure on the nerve root are not the disc itself, but the superior articular processes with their overlying ligamentum flavum.

I wish to thank Dr J. M. van Niekerk for his great help in this research project.

REFERENCES

BRAUS, H., and ELZE, C. (1954): *Anatomie des Menschen.* Band 1. Berlin: Springer-Verlag.

BROWN, T., HANSEN, R. J., and YORRA, A. J. (1957): Some Mechanical Tests on the Lumbosacral Spine with Particular Reference to the Intervertebral Discs. *Journal of Bone and Joint Surgery*, **39-A,** 1,135.

FICK, R. (1911): *Handbuch der Anatomie und Mechanik der Gelenke.* Band 3. Jena: G. Fischer.

FICK, R., and BARDELEBEN, von K. (1904): *Handbuch der Anatomie und Mechanik der Gelenke.* Band 2. Jena: G. Fischer.

FRANCESCHINI, M. (1947): Sull'architecttura collagene dei dischi intervertebrali. *Atti della Accademia delle Scienze Mediche e Naturali in Ferrara*, **26,** 1.

FRIBERG, S. (1948): Anatomical Studies on Lumbar Disc Degeneration. *Acta Orthopaedica Scandinavica*, **17,** 224.

GRANT, J. B., and SMITH, C. G. (1953): The Musculature. Section V. *Morris Human Anatomy.* Eleventh edition. New York, Toronto: The Blakiston Company.

GRAY'S ANATOMY (1958): Descriptive and Applied. Thirty-second edition. London: Longmans, Green and Co.

HAPPEY, F., HORTON, W. G., MACRAE, T. P., and NAYLOR, A. (1955): The Human Intervertebral Disk. *Nature*, **175,** 1,032.

HOAD, J. M., and ROSENBERG, P. (1953): Physics of the Intervertebral Disc, part I. *Journal of American Osteopathic Association*, **52,** 327.

HORTON, W. G. (1958): Further Observations on the Elastic Mechanism of the Intervertebral Disc. *Journal of Bone and Joint Surgery*, **40-B,** 552.

HUNTINGTON, G. S. (1902): Present Problems of Myological Research and the Significance and Classification of Muscular Variations. *American Journal of Anatomy*, **2,** 157.

LABALT, A. (1835): Quoted by Walmsley (1953).

LINDBLOM, K. (1952): Experimental Ruptures of Intervertebral Discs in Rats' Tails. *Journal of Bone and Joint Surgery*, **34-A,** 123.

LUSCHKA, H. von (1867): *Handbuch der Topographischen Anatomie, Tübingen* (Quoted by Fick 1904).

RAUBER-KOPSCH (1955): *Lehrbuch und Atlas der Anatomie des Menschen.* Band 1. von Fr. Kopsch. Stuttgart: Georg Thieme.

THIEME, F. P. (1950): Lumbar Breakdown Caused by Erect Posture in Man; with Emphasis on Spondylolisthesis and Herniated Intervertebral Disc. *Anthropological Papers, Museum of Anthropology, University of Michigan*, Paper 4.

WALMSLEY, R. (1953): The Development and Growth of the Intervertebral Disc. *Edinburgh Medical Journal*, **60,** 341.

EFFECT OF PERSISTENT PRESSURE ON THE ARTICULAR CARTILAGE

An Experimental Study

ANTONI TRIAS,* OXFORD, ENGLAND

From the Nuffield Orthopaedic Centre, Oxford

In 1951 an investigation into osteoarthritis based on clinical and experimental studies was initiated at the Nuffield Orthopaedic Centre. A number of papers have been published since 1953 reporting the progress of this work (Trueta 1953; Harrison, Schajowicz and Trueta 1953; Trueta 1954; Harrison 1955; Trueta 1956; Little and Pimm 1956; Trueta 1958; Little, Pimm and Trueta 1958; Trueta and Lima 1959). It was soon seen both in specimens taken during life and in those obtained after death that the earliest lesions of osteoarthritis were localised to the articular cartilage. This confirmed the findings of Nichols and Richardson (1909). It seemed too that the degeneration of joint cartilage resulted largely from the faulty distribution of pressures across the joint.

As part of a general plan of research into the behaviour of cartilage during growth and in osteoarthritis work was begun in 1953 on the changes produced in articular cartilage by constant heavy pressure or by intermittent pressure. A clamp similar to that designed by Charnley (1948) for arthrodesis of the knee was used to compress the knee joints of rabbits. The animals were killed after varying periods of compression and the articular cartilage and adjacent bone were examined histologically. The present work is concerned with the effect of pressure on the vascular pattern beneath the articular cartilage, and with the processes of degeneration and regeneration. Some of the findings were summarised by Trueta (1956) and further evidence was collected while the author was working in the Anatomy Department of Dalhousie University, Halifax, Nova Scotia, in 1956.

FIG. 1
Radiograph showing the compression device applied to the knee joint of a rabbit.

Some of the animals used in these experiments were also studied further for an investigation on the effects of pressure on the epiphysial growth cartilage. This work was done in collaboration with Professor Trueta and awaits publication.

MATERIAL AND METHODS

Altogether thirty-five male and female rabbits were used, all but five of them young animals. Thirty-three, of whom one was excluded because the femur broke, had the clamp introduced and pressure applied to the right knee. Two had the clamp introduced into the right knee without application of pressure. These two right knees and the left knees thus served as controls.

* An abridged account of this work was read at the meeting of the Orthopaedic Research Society, Chicago, in January 1957.

The animals were anaesthetised with intravenous nembutal and inhaled ether. Through two small longitudinal incisions over the femur and tibia two pins were introduced transversely and parallel with each other into the femur and tibia respectively at the level where the metaphysis merged with the shaft. The wounds were sutured with fine black silk. The pins were fixed by lock screws in rectangular metal blocks on both sides (Trueta 1956). Two coil springs placed between the heads of the screws and the lower blocks exerted constant pressure against the pins (Fig. 1). The springs also allowed some expansion between the two pins so as to some extent to allow for the effect of growth. The screws were tightened until a further turn would begin to bend the pins. Radiographs were then taken.

In the first three experiments external splintage was not used but it was soon noted that the rabbits tended to flex their knees. Since this meant that the same area of cartilage was not being constantly subjected to pressure, plaster immobilisation was used in all the other animals including the two controls. The knee was held fully extended by a plaster which in the early experiments extended from the groin to the toes but which in the last thirty left the foot free. The foot was bound with an elastic adhesive bandage to prevent oedema. Compression of the knee was thus maintained steadily in the vertical axis of the limb in an attempt to keep the same areas of cartilage pressed together. Nine animals were killed after compression had been maintained for varying periods. The nine were allowed to live for periods of up to two months after removal of compression and immobilisation, so that the processes of regeneration might be studied.

At a time varying from three to seventy-five days from operation the animal was anaesthetised again, and through a long median incision a cannula was inserted and tied into the abdominal aorta; through this about 70 millilitres of a 2 per cent solution of Berlin blue, or exceptionally Indian ink, were perfused. Radiographs were then taken and the animal was killed by overdose with ether. The clamps were removed and both hind limbs were disarticulated at the hip joints and fixed in 10 per cent formalin. Five to ten days later the soft tissues were removed and the knee joints disarticulated. Each bone was independently photographed and radiographed and then placed in a decalcifying solution, usually nitric acid, and finally the distal ends of the femora and the proximal ends of the tibiae were embedded in celloidin. Alternating thin (12–15μ) and thick (200–300μ) sections were cut. Most of the former were stained with haematoxylin and eosin; the thick sections were mounted on slides with thick Canada balsam and examined under the low power microscope in order to study the vessels.

The compression—The forces produced by the springs were estimated. At their maximum, towards the end of the experiment, the compression force exerted by each spring varied from 3·5 to 4·5 kilograms. The total compressing force across the joint was, therefore, of the order of 7 to 9 kilograms (from 15 to 20 pounds).

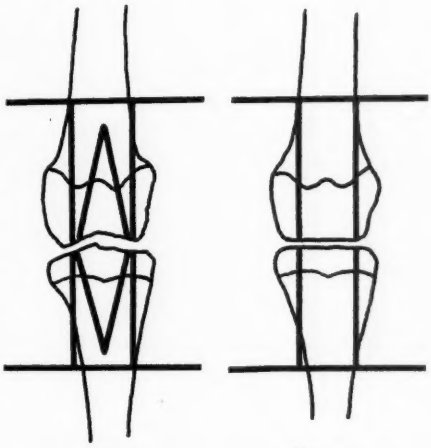


FIG. 2 FIG. 3

Figure 2—The lines of compression take the shape of four inverted cones. Figure 3—Showing the conversion to cylindrical area of compression.

The area of contact between the femur and tibia was estimated at one-third of a square centimetre and the total pressure per square centimetre between the two bones was considered to be from 21 to 27 kilograms (289 to 371 pounds per square inch).

It is clear that the lines of compression were directed from the pins towards the small areas of cartilage contact, in the shape of four cones, two in each bone with a common base

and two apices in the area of joint contact (Fig. 2). In this diagram the four cones are shown as they act at the beginning of the experiment. Later, with progressive flattening of the area of contact, this becomes larger and the four cones of compression tend to merge into one single cylindrical column (Fig. 3). This change decreases the amount of pressure per square centimetre. Fifteen animals had the pressure exerted constantly on the knee for varying periods. Fourteen had the pressure applied and released during alternate periods of one hour for eight hours each day until they were killed from four to twelve days later.

FIG. 4
Showing lack of staining in the zone of cartilage subjected to compression. (×15.)

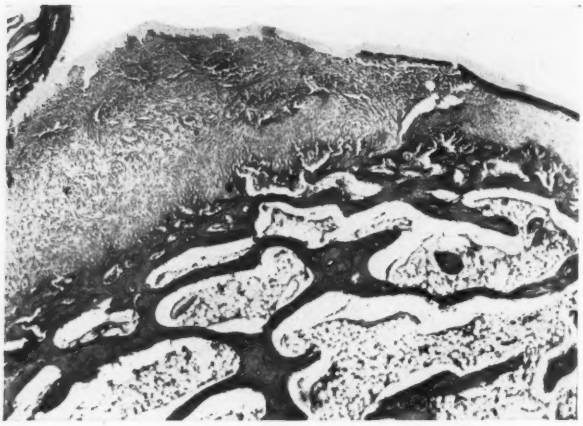


FIG. 5
The degenerate fibrous tissue breaks away from the remaining cartilage. (×15.)

RESULTS

The efficiency of immobilisation—It soon became apparent that immobilisation of the knee joint by a plaster cast from foot to groin still allowed some flexion of the joint, which would tend to displace the area of maximum pressure over the articular cartilage from the front to the back of the joint. In almost all the animals examined, this moderate but clear tendency to flexion was detectable by signs of regeneration of cartilage.

In all cases the radiographs taken after compression showed the expected reduction of "joint space."

The effect of constant pressure—After four days of constant pressure the articular cartilage was found to be severely affected. The changes were confined to the areas of direct contact between the joint cartilages of the tibia and femur. The first histological evidence of damage was the loss of nuclear staining of the chondrocytes although their boundaries were still visible. The cartilage kept its normal width but did not stain like that outside the area of

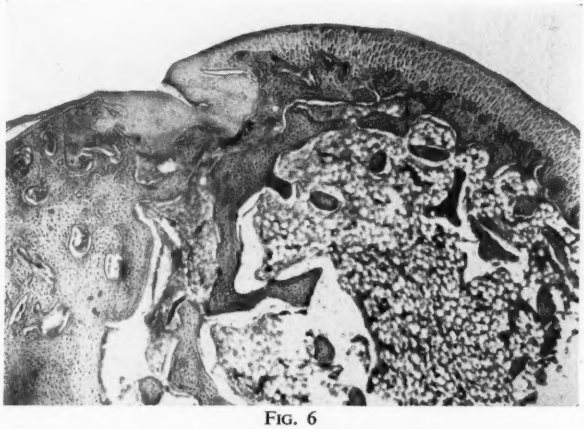


FIG. 6

The bone at the left side of this section is dense, corresponding to the area subjected to excessive pressure. On the right, under the regenerated cartilage, bone resorption has occurred. (×15.)

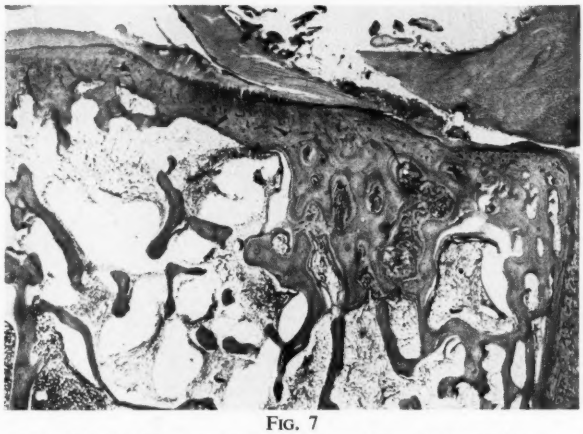


FIG. 7

On the left of this section, bone atrophy underneath the area free from compression is seen. On the right, under the necrotic tissue, a large area of bone sclerosis occurs. (×6.)

maximum pressure (Fig. 4). The superficial layer of the cartilage was the first to show this change, but when compression was maintained for longer periods the paler staining affected the cells of the deeper part of the cartilage also, eventually involving the whole thickness, layer after layer.

The next sign of cartilage damage was its progressive thinning and the disappearance of the cell boundaries of the chondrocytes. At this stage the cartilage became fibrillated.

At the next and last stage of degeneration it was found that the vessels penetrated the cartilage from the underlying epiphysial bone and substitution by fibrous tissue occurred. This appeared disorganised and detached from the epiphysis despite the lack of any appreciable movement in the joint, apart from the very limited tendency of the joint to flex (Fig. 5).

Changes in the zone of calcification and underlying bone—Underneath the compressed cartilage changes followed a similar pattern, being first detectable in the superficial area of calcified cartilage and progressively invading the deep bone structures to change the orientation and thickness of the trabeculae. In its early stages the zone of calcification underneath the compressed cartilage was seen to have increased in thickness. Instead of its normal regular shape and narrow width the area of calcification became wide, merging into the underlying trabeculae which appeared irregular and much increased in thickness, not only in the epiphysis but also in the metaphysis (Fig. 6). In contrast with this augmented bone density in the area of excessive pressure, the non-compressed segment of the epiphysis appeared to be almost deprived of trabeculae (Fig. 7).

In most of the animals in which pressure was maintained for up to twelve days it was repeatedly seen that lamellae in the compressed area had lost their osteocytes and that new lamellae

FIG. 8
New lamellar bone being laid down over dead trabeculae. (×75.)

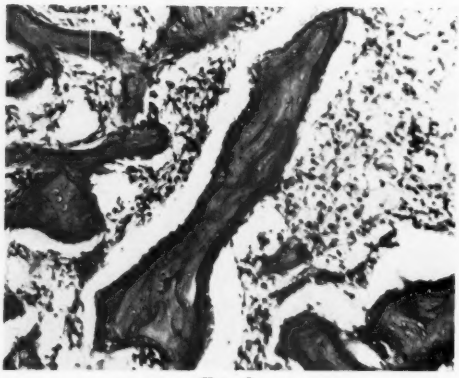


FIG. 9
New bone being formed over trabeculae which still mostly survive. (×65.)

FIG. 10

FIG. 10
A profusion of large vessels occupies the bone marrow of the epiphysis under the dead cartilage. (×10.)

appeared covering the dead ones, thus contributing to the total increase of the bone thickness in the area of excessive pressure (Fig. 8). On other occasions active new bone formation was seen to take place over apparently viable trabeculae (Fig. 9). In thick sections large vessels were seen occupying the marrow spaces along and between the thickened trabeculae (Fig. 10).

The effect of constant heavy pressure—In all the rabbits in which the compression clamp was left on the knee joint for more than twelve days, the decrease in the subjacent marrow spaces was such that vessels became scarce. Under these circumstances a new phase of bone death occurred, apparently due to the excessive thickness of the bone and consequent reduction of blood flow for lack of bone marrow spaces. In this way round areas of fibrous tissue and debris, deprived of vessels, occurred; they were suggestive of the bone "cysts" associated with advanced osteoarthritis in man (Figs. 11 to 14).

Effect of intermittent pressure—Fourteen rabbits were submitted to the same experimental conditions as the group described above. The only variant was that in eleven animals daily pressure was applied on and off, one hour at a time, for four hours. The animals were killed at different times from four to twelve days. Two animals had twelve hours on and twelve hours off pressure on alternate days. They were subsequently killed at varying times. One animal was discarded because of femoral fracture.

It was noted that even thirty-seven days after the removal of the clamps the rabbit which had had its knee joint compressed for twelve hours on alternate days, when killed twelve days later, showed advanced degeneration of cartilage with areas of severe damage with eburnation (Fig. 15). After seventeen days of constant compression, an animal allowed to survive for a

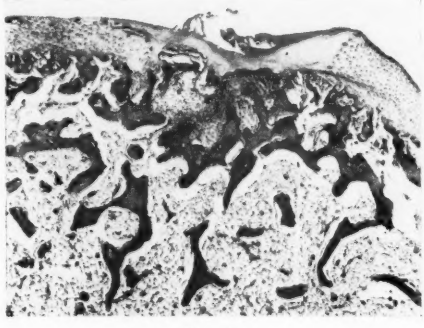


FIG. 11

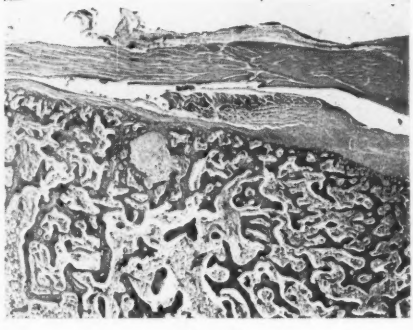


FIG. 12

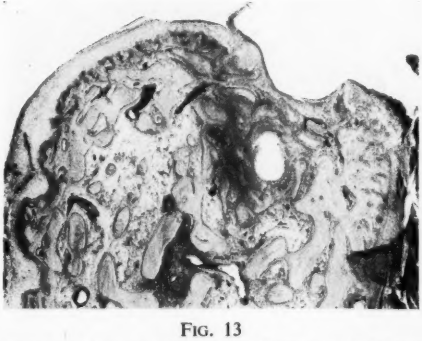


FIG. 13

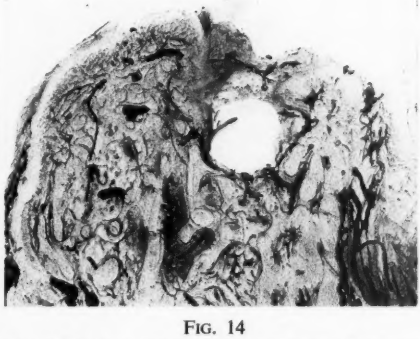


FIG. 14

Process of " cyst " formation. Figure 11—Degeneration of the bone under the area of greatest pressure when cartilage protection is gone. (×13.) Figure 12—A "cyst" appears always "open" to the joint cavity. (×10.) Figure 13—The " cyst " is now half empty. (×6.) Figure 14—Vessels surround the " cyst " but do not penetrate into it. (×10.)

period of seventy-five days showed advanced degeneration of cartilage but at the same time remarkable signs of regeneration. Other animals left to survive after continuous pressure for twelve days or more showed very moderate if any regeneration (Fig. 16).

None of the eleven rabbits in which intermittent joint pressure had been applied for four hours daily showed signs of severe cartilage degeneration and in two of them no signs of degeneration whatsoever were noted.

Processes of regeneration—The earliest sign of regeneration of cartilage was manifested by the increased mitotic activity of the chondrocytes and their heavy staining. It was noted that the deep chondrocytes tended to remain alive much longer than the superficial ones and in their reproductive activity the deep cartilage cells surpassed those lying more superficially (Fig. 17). Even in those animals in which the superficial part of the cartilage was completely disintegrated the reparative activity of the more deeply situated chondrocytes was often seen (Fig. 18), mitosis occurring most frequently in the deepest layers of the cartilage.

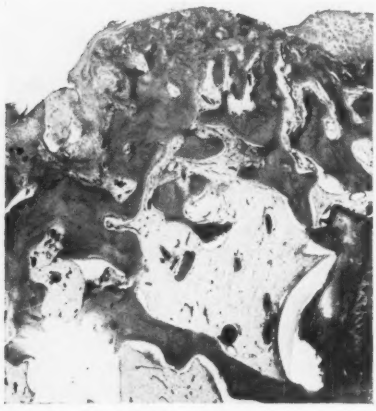


FIG. 15

FIG. 16

Figure 15—Large area of eburnation. (×14.) Figure 16—Moderate regeneration of cartilage seventy-five days after release of compression. (×14.)

It was apparent that the severely damaged cartilage with all its cells dead and its collagen fibres disintegrated had no capacity for regeneration. Even two months after the removal of a twelve-day constant compression, this part of the cartilage remained dead and in the process of elimination into the joint cavity. In these cases mitotic activity was seen in the chondrocytes adjacent to the dead area, but even as late as sixty days after removal of compression they had not invaded the dead area.

In some animals, as long as fifty days after removal of constant compression for more than twelve days, the area of transition between the dead and the living cartilage was marked by a straight line at 90 degrees with the joint surface (Fig. 19). This vertical alignment of the dividing line was a clear indication of the limits of the cones of pressure we have mentioned above.

In active regeneration clusters of chondrocytes appeared surrounded by an apparently new capsule. Their number varied, frequently far surpassing the normal two to eight. The matrix was also more abundant around the chondrocytes.

Proliferation of young connective tissue occurred in the bone marrow. It was most striking to observe the frequency with which new chondrocytes evolved from this invading connective tissue. It was also interesting to note that the vessels initially surrounded by the young connective tissue disappeared as soon as the superficial layer of the fibrous articular cartilage was restored (Fig. 20).

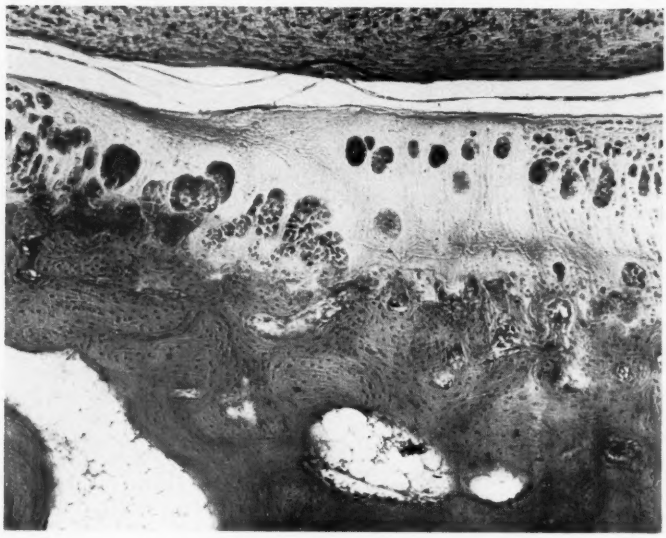


FIG. 17
Deep nests of chondroblasts showing proliferative activity; the superficial layers of cartilage are dead. (×45.)

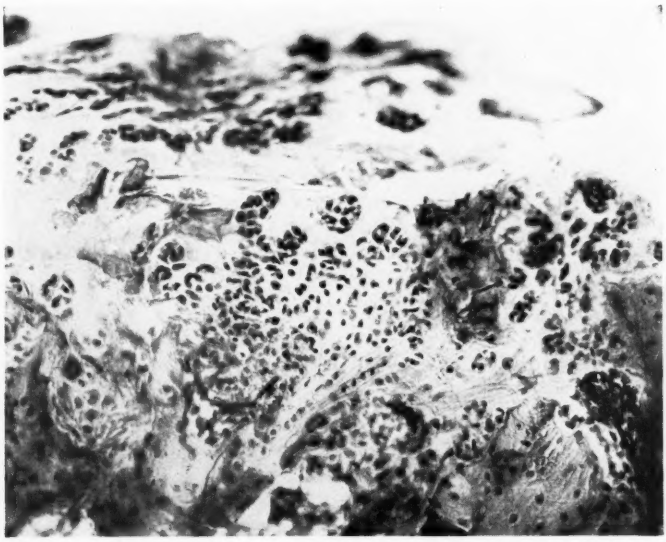


FIG. 18
New clusters of chondrocytes appear in the deep area of fibrous degeneration connected with the bone marrow. (×60.)

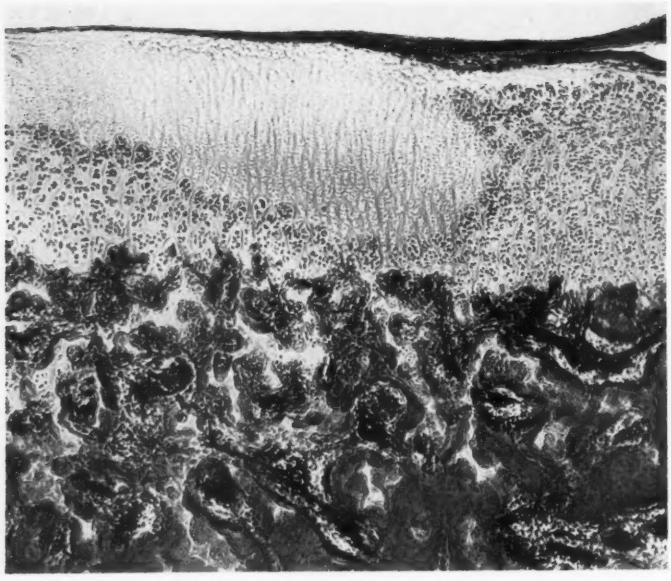


FIG. 19
Sharp line of separation between living and dead chondrocytes. (×24.)

FIG. 20
Restoration of fibrous articular cartilage and cessation of vascular penetration. (×24.)

DISCUSSION

Many workers have attempted to reproduce degenerative arthritis experimentally in order to study the natural history of the disease as it occurs in man. Axhausen (1912) destroyed the joint cartilage with chemicals or with a cautery, and postulated that the arthritic changes were due to the pressure of dead cartilage within the joint. In repeating these experiments, Burckhardt (1924) thought that degenerative arthritis was a reactive phenomenon due to cartilage injury. That view was also supported by Fisher (1922) who caused proliferative changes on the margins of the joint cartilage after creating a defect in the joint cartilage. On the other hand, Key (1931) failed to reproduce similar changes with the same technique. Bennett and Bauer (1937) succeeded in causing degeneration-regeneration, producing defects in the patella, and Carlson (1957) confirmed these findings. These and many other experimental studies of degenerative arthritis aimed at causing direct damage to the joint cartilage, to elicit degenerative changes. However, it is well recognised that degenerative arthritis in man appears frequently in joints which have been free from any direct damage, either traumatic or inflammatory.

A new line of approach has come along since the recent studies on the nature of cartilage matrix. We now know that it is constituted of collagen fibres, water and mucopolysaccharides, mainly chondroitin sulphate. The chondroblast is now considered responsible for laying down both the collagen and chondroitin sulphate. It is thus apparent that for the cartilage to degenerate, either the chondrocyte must die from direct injury, as in Axhausen's experiments, or else its metabolism must be seriously impaired. The work of Harrison, Schajowicz and Trueta (1953) firmly supports these views.

Reinhardt and Li (1953) showed that the administration of growth hormones causes arthritic changes and Ponseti and Shephard (1954) reported that poisoning animals with nitriles produced lesions in all tissues containing chondroitin sulphate by altering the metabolism of the hexosamines in this complex.

Contrary to the belief prevailing not long ago, it is now recognised since the work of Holmdahl and Ingelmark (1950), and that of Trueta and co-workers, that while the cartilage is devoid of blood vessels, it receives its nourishment by diffusion and imbibition of the surrounding fluids. These come not only from the vessels of the synovial membrane, which are mainly responsible for the formation of synovial fluid, but also from the vessels under the calcified cartilage of the bone epiphysis. Immobilisation of the joint interferes with the normal mechanisms of fluid transport, one of which—intermittent compression of the cartilage during movement—has been shown to be important in bringing nutritive joint fluids to the chondrocytes. Evans, Eggers, Butler and Blumel (1960) have shown that lack of normal movements may in fact lead to degenerative changes in joint cartilage. Harrison and his colleagues (1953) have shown that in degenerative arthritis in man the earliest lesion is found in the non-weight-bearing areas of the joint which are not subjected to the pumping action that occurs in the weight-bearing area.

This evidence seemed to indicate that a study of the effects of various forms of compression on the joint cartilage might be rewarding. An experimental work based on some ideas similar to these has just been published by Salter and Field (1960). In their paper they state that " no reference to a previous study of this aspect of articular cartilage could be found in the literature." They have overlooked the publications by Trueta (1956, 1958) which gave a preliminary account of the present work.

SUMMARY

The effects on articular cartilage of continuous and intermittent excessive pressures have been studied in the knees of rabbits. Severe degenerative changes in the cartilage were observed; these resembled the typical lesions seen in osteoarthritis in man. They included fibrillation of cartilage, death of chondrocytes, eburnation of joint surfaces, sclerosis of bone and the

production of " bone cysts." Regeneration of cartilage was common and it was brought about either by the deeply situated chondrocytes which had escaped death or by metaplasia of young connective tissue cells of the bone marrow.

I am indebted to Professor J. Trueta for the planning of this work and his help and inspiration throughout. I also want to thank Professor R. L. de C. H. Saunders and Dr F. W. Fyfe for their help and constructive criticism, and Dr Carl Saiphoo for his technical assistance.

The work reported in this paper was carried out while the author held the Girdlestone Memorial Scholarship in the autumn of 1954. Part of it was supported by two grants from the Canadian National Research Council.

REFERENCES

AXHAUSEN, G. (1912): Ueber einfache, aseptische Knochen- und Knorpelnekrose, Chondritis dissecans und Arthritis deformans. *Archiv für Klinische Chirurgie*, **99,** 519.

BENNETT, G. A., and BAUER, W. (1937): Joint Changes Resulting from Patellar Displacement and their Relation to Degenerative Joint Disease. *Journal of Bone and Joint Surgery*, **19,** 667.

BURCKHARDT, H. (1924): Experimentelle Untersuchungen über die Beziehungen der Gelenkfunktion zur Arthritis deformans. *Archiv für Klinische Chirurgie*, **132,** 706.

CARLSON, H. (1957): Reactions of Rabbit Patellar Cartilage Following Operative Defects. A Morphological and Autoradiographic Study. *Acta Orthopaedica Scandinavica*, Supplementum 28.

CHARNLEY, J. C. (1948): Positive Pressure in Arthrodesis of the Knee Joint. *Journal of Bone and Joint Surgery*, **30–B,** 478.

EVANS, E. B., EGGERS, G. W. N., BUTLER, J. K., and BLUMEL, J. (1960): Experimental Immobilization and Remobilization of Rat Knee Joints. *Journal of Bone and Joint Surgery*, **42–A,** 737.

FISHER, A. G. T. (1922): A Contribution to the Pathology and Etiology of Osteoarthritis; with Observations upon the Principles Underlying its Surgical Treatment. *British Journal of Surgery*, **10,** 52.

HARRISON, M. H. M. (1955): Present Trends in the Treatment of Osteoarthritis of the Hip. *Postgraduate Medical Journal*, **31,** 397.

HARRISON, M. H. M., SCHAJOWICZ, F., and TRUETA, J. (1953): Osteoarthritis of the Hip: A Study of the Nature and Evolution of the Disease. *Journal of Bone and Joint Surgery*, **35–B,** 598.

HOLMDAHL, D. E., and INGELMARK, B. E. (1950): The Contact Between Articular Cartilage and Medullary Cavities of the Bone. *Acta Orthopaedica Scandinavica*, **20,** 156.

KEY, J. A. (1931): Experimental Arthritis; the Changes in Joints Produced by Creating Defects in the Articular Cartilage. *Journal of Bone and Joint Surgery*, **13,** 725.

LITTLE, K., and PIMM, L. H. (1956): Osteoarthritis of the Hip Joint. *Proceedings Stockholm Conference on Electron Microscopy*, 233.

LITTLE, K., PIMM, L. H., and TRUETA, J. (1958): Osteoarthritis of the Hip. *Journal of Bone and Joint Surgery*, **40–B,** 123.

NICHOLS, E. H., and RICHARDSON, F. L. (1909): Arthritis deformans. *Journal of Medical Research*, **21,** 149.

PONSETI, I. V., and SHEPHARD, R. S. (1954): Lesions of the Skeleton and of other Mesodermal Tissues in Rats Fed Sweet-pea (*Lathyrus Odoratus*) Seeds. *Journal of Bone and Joint Surgery*, **36–A,** 1,031.

REINHARDT, W. O., and LI, C. H. (1953): Experimental Production of Arthritis in Rats by Hypophyseal Growth Hormone. *Science*, **117,** 295.

SALTER, R. B., and FIELD, P. (1960): The Effects of Continuous Compression on Living Articular Cartilage: an Experimental Investigation. *Journal of Bone and Joint Surgery*, **42–A,** 31.

TRUETA, J. (1953): Recherches récentes sur la pathogénie de la coxarthrose. *Mémoires de l'Académie de Chirurgie*, **79,** 427.

TRUETA, J. (1954): Osteoarthritis of the Hip. *Annals of the Royal College of Surgeons of England*, **15,** 174.

TRUETA, J. (1956): Osteo-arthritis; an Approach to Surgical Treatment. *Lancet*, **i,** 585.

TRUETA, J. (1958): Considerations of the Pathology of Osteoarthritis of the Hip. *Société Internationale de Chirurgie Orthopédique et de Traumatologie. Septième Congrès International de Chirurgie Orthopédique Barcelone*, 16–21 *Septembre* 1957. Procès-verbaux. Bruxelles: Imprimerie des Sciences.

TRUETA, J., and LIMA, C. (1959): Ageing and Osteoarthritis. *Gerontologia Clinica*, **1,** 283.

WAWZONEK, S., PONSETI, I. V., SHEPHARD, R. S., and WIEDENMANN, L. G. (1955): Epiphyseal Plate Lesions, Degenerative Arthritis, and Dissecting Aneurysm of Aorta Produced by Aminonitriles. *Science*, **121,** 63.

GENETICALLY CONTROLLED DEGENERATION OF THE NUCLEUS PULPOSUS IN THE MOUSE

R. J. BERRY, LONDON, ENGLAND

From the Medical Research Council Experimental Genetics Research Unit, University College, London

In the past few years ideas on the etiology of intervertebral disc protrusion have undergone a fairly radical change, and " it is now generally conceded that trauma is not a large factor in the production of disc prolapse, and may often be absent " (Annotation, *Lancet* 1958). As long ago as 1948 Jackson suggested that there are anatomical defects which predispose to disc protrusion and which are " most likely of a developmental nature, possibly increased by other levers in the form of trauma, toxaemia and pregnancy." Histochemical and biophysical studies of the discs (Charnley 1952, Malinský 1957, Hendry 1958, Naylor 1958) seem to point to the conclusion that predisposing weaknesses in the discs may be regarded as similar to normal ageing changes. However, " gross degeneration in older people is rarely associated with prolapse. If desiccation in discs is a major factor in their prolapse it is difficult to understand why the elderly remain relatively free from major disc derangements " (Annotation, *Lancet* 1958). This fact must remain unexplained unless it can be shown that certain people have an inherent proneness to disc lesions, or, in other words, unless it is found possible to divide the population into those whose discs show such changes during the first twenty or thirty years of life that they are predisposed to prolapse, and those whose discs undergo only "normal" changes during life.

However, just such changes have been shown in at least two experimental animals. Hansen (1951, 1952) showed that markedly different rates of disc prolapse exist in different breeds of dogs, and that prolapse in these animals is certainly due to degenerative changes in the nucleus pulposus. The purpose of this communication is to describe a similar genetically determined effect in the house mouse, resulting from a progressive reduction of the notochord in a cephalo-caudad direction (Berry 1960).

THE PINTAIL MUTATION IN THE MOUSE

The work described in this paper refers to a mutation which was found in a mouse of the pBr strain which had been treated with the powerful carcinogen—and alleged mouse mutagen—methylcholanthrene (Hollander and Strong 1951). The tail of this mouse was about " two-thirds normal length, and the distal end showed a peculiar thin and somewhat distorted tip." The name " Pintail " (symbol *Pt*) was given to this trait. The gene normally behaves as a partial dominant, and mice heterozygous (*Pt*/+) and homozygous (*Pt*/*Pt*) for the gene are fertile. Homozygous Pintail mice have hardly any tail and are often debilitated; they frequently have intestinal abnormalities.

Pintail mice usually have one or more kinks in the distal part of the tail (Fig. 2), but several Pintail heterozygotes have occurred with no tail kinking. The probable explanation is that the tail kinks are the incidental result of abnormal intervertebral discs which are insufficient to maintain the vertebrae in their correct relationship to one another. This conclusion is strengthened by the observation that tail kinks only appear at about the age of two weeks, at which time the mice leave the nest and run around actively.

THE EFFECT OF THE PINTAIL GENE ON THE INTERVERTEBRAL DISCS

The Pintail gene has an effect on the intervertebral discs which is not obvious from an examination of the living animals, but which is only revealed by histological techniques.

One of these techniques must be described. Grüneberg (1958) in a personal communication observed that the mucoid nucleus pulposus is stained very clearly in methylene blue clearance preparations (Noback 1916, Grüneberg 1953). This observation was utilised and the axial skeleton of a number of normal and abnormal animals aged three weeks and older was stained

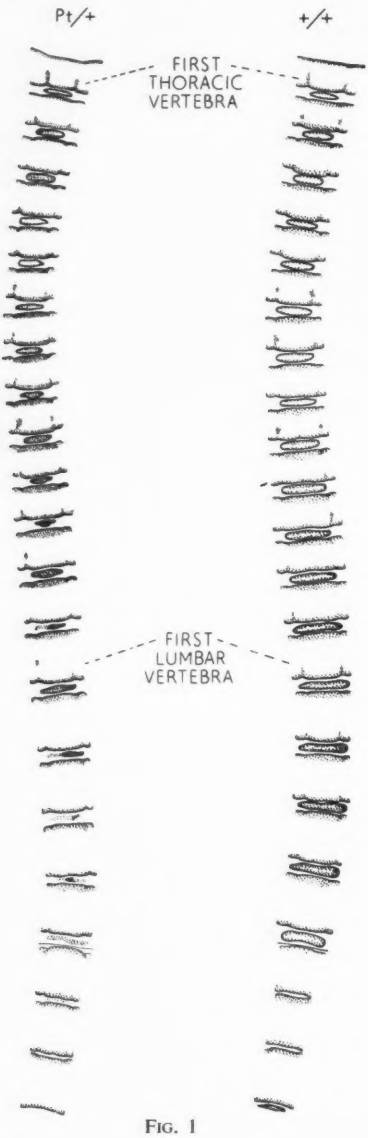


FIG. 1

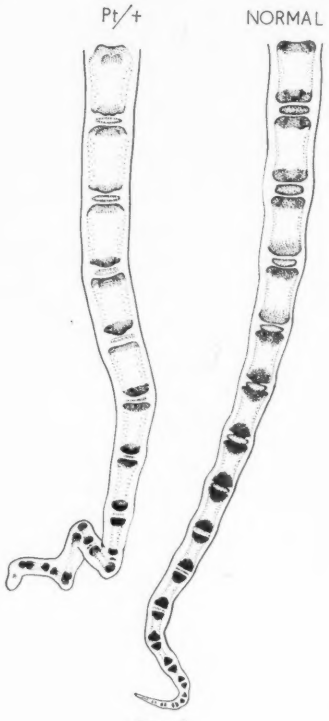


FIG. 2

Figure 1—Freehand drawings of methylene blue stained clearance preparations of the vertebral column of *Pt*/+ (*left*) and +/+ (*right*) litter mates at twenty-five days old to show the variation in size and shape of the nuclei pulposi.

Figure 2—Freehand drawings of the distal two-thirds of the tails of mice from the same litter as those illustrated in Figure 1. (Reproduced from *Genetical Research*, **1**, 1960.)

to show the nucleus pulposus. The animals were killed and the vertebral column removed, straightened and tied to a glass plate, and fixed in formol saline. The specimens were stained with methylene blue, dehydrated with alcohol and cleared in methyl salicylate. The best preparations are those of animals aged about three weeks. Soon afterwards the affinity of

the nuclei pulposi in the trunk region for the stain diminishes. This decrease in staining property is rather quicker in Pintails than in normals of the same age, but by the age of eight weeks or so the discs in the cervical region of both genotypes stain hardly at all.

There are differences in both the size and staining intensity of the nuclei pulposi in normals and Pintails (Figs. 1 and 2). The nuclei pulposi in the cervical region are more or less normal. More caudad the size of the nucleus pulposus progressively decreases. The nuclei pulposi in the normal mouse are more or less evenly stained the whole way down the vertebral column; in the Pintail the staining becomes more intense as the discs are reduced in size. This increase in staining intensity is accompanied by a decrease in the definition of the staining boundary of each nucleus pulposus, particularly in the tail, and the shape also changes in a characteristic fashion from head to tail. Between the cervical vertebrae in both normal and Pintail mice the nuclei pulposi are roughly cigar shaped as seen in a thoraco-ventral projection, in the thoracic region those of Pintails are more spherical than those of normals, and in the lumbar region they are flatter and thinner—but the outlines of some of those more caudad are not completely regular. The nuclei pulposi in normal mice reach their greatest size in the lower lumbar region. There are no nuclei pulposi between the first three sacral vertebrae.

MEASUREMENT OF THE NUCLEI PULPOSI

In an attempt to make the above observations quantitative, the two main diameters of the nucleus pulposus in clearance preparations were measured under a microscope fitted with

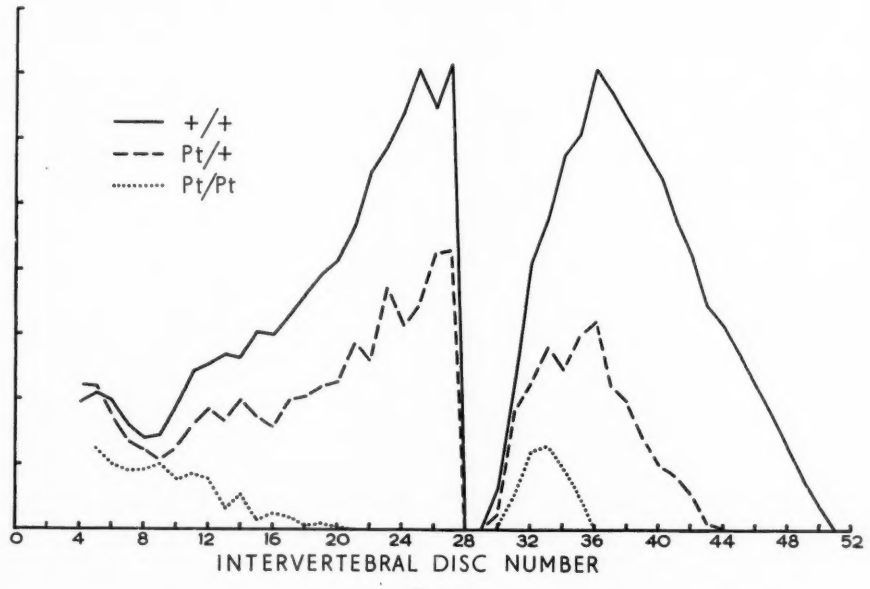


FIG. 3

Variation in size of the nucleus pulposus along the vertebral column. The vertical ordinate is the product of the two main diameters in arbitrary units. Mean values of a litter of two +/+ (continuous line), three *Pt*/+ (broken line) and one *Pt*/*Pt* (dotted line) animals aged twenty-one days.

a Leitz micrometer screw objective. It was possible to measure these dimensions for almost the whole of the length of the vertebral column, although the diffuse boundary of the staining in the last few tail discs made accurate measurement difficult. Measurements were made upon a series of litters containing normal, *Pt*/+, and *Pt*/*Pt* animals. All three genotypes

had a fairly constant pattern of size variation along the column, although there was quite a lot of both inter- and intra-litter variation. The product of the two diameters was found best to characterise each nucleus pulposus. The sizes of the nuclei pulposi in all the genotypes

FIG. 4

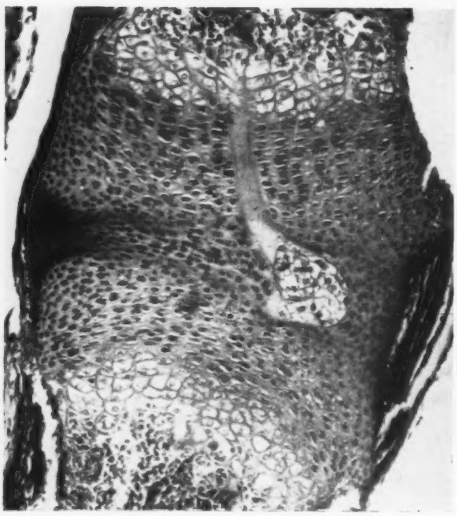


FIG. 5

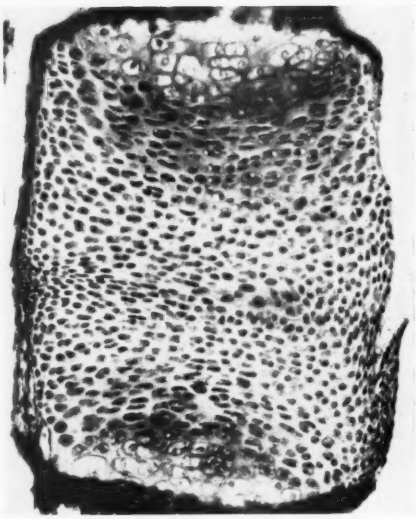


FIG. 6

FIGS. 4 TO 6

Photomicrographs of sagittal 10μ sections through lumbar intervertebral discs stained with alcian blue and chlorantine fast red of newborn +/+ (Fig. 4), *Pt*/+ (Fig. 5) and *Pt*/*Pt* (Fig. 6) litter mates. (×115.)

were of the same order in the cervical region; from about the tenth disc caudad, the abnormals showed a marked reduction in size; in the Pintail homozygote there were virtually no nuclei pulposi in the posterior thoracic and lumbar regions. The progressive nature of the reduction in size along the vertebral column can be clearly seen in Figure 3.

HISTOLOGICAL CHANGES

The consequences of this reduction in size of the nuclei pulposi were investigated histologically. Because of its medical interest, the lumbo-sacral section of the spine was studied in preference to others. The material was fixed in Bouin's fluid, decalcified with 2 per cent nitric acid, embedded by Peterfi's technique and serially sectioned at 10μ in the sagittal plane. The sections were stained with Ehrlich's haematoxylin and eosin, or alcian blue and chlorantine fast red (Lison 1954).

At birth (Figs. 4 to 6) the nucleus pulposus was little differentiated; in both normal and *Pt*/+ mice it consisted of simple aggregations of notochordal cells, which were well stained and whose cell wall reticulum was unbroken. The first signs of acid mucopolysaccharides (stained with alcian blue) penetrating into the cellular structure were apparent, and this process seemed to have advanced farther in the abnormal than in the normal. In conformity

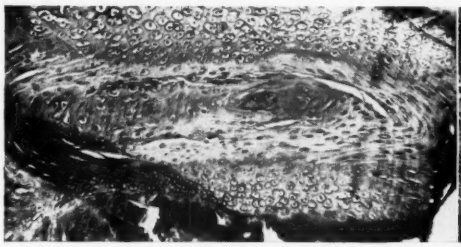


FIG. 7

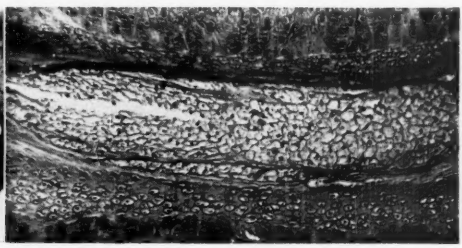


FIG. 8

Similar sections to those shown in Figures 4 to 6 to show the more advanced changes in the nucleus pulposus of the abnormal in fifty-day-old *Pt*/+ (Fig. 7) and +/+ (Fig. 8) litter mates. (×80.)

with the findings in clearance preparations there was no sign of any nucleus pulposus in the *Pt*/*Pt* animal, though traces of the notochordal sheath were found in some sections. The actual disc region consisted mainly of cartilaginous tissue with much acid mucopolysaccharide content, but the tissue was freely interspersed with collagen fibres (stained red with chlorantine fast red).

The penetration of acid mucopolysaccharides into the nucleus pulposus proceeded rapidly during early post-natal life. Already at five days it was clearly surrounded by a zone of mucoid material containing no nuclei, and the penetration of this substance between the cells of the primitive nucleus pulposus had been sufficient to isolate islands of cells. The cell nuclei in *Pt*/+ are enlarged and stained less well than at birth. By ten days the whole of the small cellular nucleus pulposus of *Pt*/+ was divided into blocks surrounded by homogeneous mucoid substance, and there was more collagen in the surrounding regions.

No trace of the original notochordal cell structure was left in a thirty-nine-day-old *Pt*/+. The central disc area stained badly with haematoxylin and eosin, but seemed to contain many fibrocytes. A definite central nucleus pulposus existed at fifty days (Fig. 7). This area was almost entirely mucoid with many large nuclei. It was not clear whether these were the remains of notochordal cells, or were fibrocytes and cartilage cells that had immigrated from outside. There was no sign of the original cell wall reticulum. In the normal at this age Fig. 8) the original cell walls were still present and the nuclei clearly visible, and the most obvious change from the early post-natal structure was in the continuing ingression of the cellular area, with some cells seeming to have been confined in vacuoles in the mucoid substance, and the nuclei of these cells showed some signs of disintegration. The annulus fibrosus of both normal and Pintail discs seemed to differentiate normally.

Between fifty and a hundred days the mucoid structure of the *Pt*/+ disc was largely infiltrated with collagen. Ossification of parts of the annulus fibrosus on the inside of the vertebral end-plates sometimes occurred.

There were no further major changes. At 250 days the *Pt*/+ disc was more fibrous than formerly, while the cell wall reticulum of the normal was still clearly visible, although the nuclei were now only just discernible. In fact the lumbar discs of *Pt*/+ animals changed more in the first seven weeks after birth than did those of normals in the first seven months.

DISCUSSION

The effect of the Pintail gene on development has been shown (Berry 1960) to be a reduction in the mitotic rate in the notochord beginning on the tenth day of gestation (birth takes place on the nineteenth or twentieth day). The only essential difference between newborn normal and Pintail mice is that there are more notochordal cells in the former. Evidently the greatly accelerated age changes in the discs of Pintail mice are a direct consequence of their reduced size.

Peacock (1952) and Malinský (1959) have described the changes that take place in the discs of man with increasing age. The main respect in which they differ from those in the mouse is in the more rapid disappearance of notochordal cells. The work of Hansen (1951, 1952) on the different dispositions of certain breeds of dog to disc degeneration and prolapse has already been mentioned. The age changes in the discs of breeds subject to disc degeneration closely resemble those in Pintail mice. This condition is a breed characteristic in dogs and hence presumably genetic though it is uncertain whether it is produced by a single gene as is Pintail in the mouse.

Many authors have studied the changes in protruded discs in man. Eckert and Decker (1947) found differences between discs removed at operation and those taken from cadavers of similar ages, the most striking being stroma degeneration of the nucleus pulposus in the operatively removed discs. Hendry (1958) showed that protruded discs in man have a lowered capacity to take up water; he ascribed this to the " loss of or deterioration in the muco-protein gel of the nucleus—that is, the same changes as have been shown by Sylvén (1951) and others to occur normally only with advancing years." Mitchell, Hendry and Billewicz (1961) have shown that the polysaccharide content of a series of prolapsed discs removed at operation was " strikingly uniform " and lower than that of a comparable series of normal discs, in which the polysaccharide content varies with age, declining from the fourth decade onwards. Mice which carry the *Pt* gene show just such a loss of muco-protein in the nucleus pulposus, that is, they exhibit a genetically determined weakness of the intervertebral discs.

The suggestion put forward in this paper is that disc protrusion often, if not always, results from a congenital weakness of the discs themselves. The direct proof of the genetic basis of such a weakness in man would be very difficult: a predisposition to disc protrusion in the third or fourth decade of life is not a subject which lends itself easily to family studies, particularly since there are sex and occupational differences in the incidence of the condition. There may be a gene similar to Pintail widespread in human populations. It would presumably manifest itself in no other way than giving its possessors a " weak back." However, a genetical hypothesis answers the difficulty expressed by the *Lancet* annotation (1958), that " it is difficult to understand why the elderly remain relatively free from major disc derangements." If there is a human gene which causes accelerated ageing changes, carriers of it would be particularly liable to disc lesions comparatively early in life. People without such a gene for premature ageing would reach a similar degree of disc degeneration only at an age when their relative immobility would tend to protect them from injury.

SUMMARY

A description is given of a mutation in the mouse which reduces the size of the nucleus pulposus in the intervertebral disc of the adults. The discs of these mice show greatly accelerated age changes. The consequences of a similar mutation in man are discussed.

I am indebted to Professor Hans Grüneberg, F.R.S., for his constant encouragement and criticism, to Miss June Denny for the photography, and to Mr A. J. Lee for the drawings. The work was supported by a grant from the Medical Research Council which is gratefully acknowledged.

REFERENCES

BERRY, R. J. (1960): Genetical Studies on the Skeleton of the Mouse. XXVI. Pintail. *Genetical Research*, **1**, 439.

CHARNLEY, J. (1952): The Imbibition of Fluid as a Cause of Herniation of the Nucleus Pulposus. *Lancet*, **i**, 124.

ECKERT, C., and DECKER, A. (1947): Pathological Studies of Intervertebral Discs. *Journal of Bone and Joint Surgery*, **29**, 447.

GRÜNEBERG, H. (1953): Genetical Studies on the Skeleton of the Mouse. VII. Congenital Hydrocephalus. *Journal of Genetics*, **51**, 327.

GRÜNEBERG, H. (1958): *Personal communication.*

HANSEN, H.-J. (1951): A Pathologic-anatomical Interpretation of Disc Degeneration in Dogs. *Acta Orthopaedica Scandinavica*, **20**, 280.

HANSEN, H.-J. (1952): A Pathological-anatomical Study of Disc Degeneration in Dog. *Acta Orthopaedica Scandinavica*, Supplementum **11.**

HENDRY, N. G. C. (1958): The Hydration of the Nucleus Pulposus and its Relation to Intervertebral Disc Derangement. *Journal of Bone and Joint Surgery*, **40–B**, 132.

HOLLANDER, W. F., and STRONG, L. C. (1951): Pintail, a Dominant Mutation Linked with Brown in the House Mouse. *Journal of Heredity*, **42**, 179.

JACKSON, H. (1948): The Association Between Certain Anatomical Facts, Normal and Morbid, and the Symptomatology of Intervertebral Disc Protrusions in the Lumbar Region. *Annals of Royal College of Surgeons of England*, **2**, 273.

LANCET (1958): Annotation: Physical Changes in the Prolapsed Disc, **i**, 1,214.

LISON, L. (1954): Alcian Blue 8G with Chlorantine Fast Red 5B. A Technic for Selective Staining of Mucopolysaccharides. *Stain Technology*, **29**, 131.

MALINSKÝ, J. (1957): Histochemical Demonstration of Carbohydrates in Human Intervertebral Discs During Postnatal Development. *Acta Histochemica*, **5**, 120.

MALINSKÝ, J. (1959): Histochemistry of Carbohydrates and Lipids in Intervertebral Discs During Phylogenetic Development. *Acta Histochemica*, **7**, 107.

MITCHELL, P. E. G., HENDRY, N. G. C., and BILLEWICZ, W. Z. (1961): The Chemical Background to Intervertebral Disc Prolapse. *Journal of Bone and Joint Surgery*, **43–B**, 141.

NAYLOR, A. (1958): Changes in the Human Intervertebral Disc with Age. *Proceedings of Royal Society of Medicine* (Section of Comparative Medicine), **51**, 573.

NOBACK, G. J. (1916): The Use of the Van Wijhe Method for the Staining of the Cartilaginous Skeleton. *Anatomical Record*, **11**, 292.

PEACOCK, A. (1952): Observations on the Post-natal Structure of the Intervertebral Disc in Man. *Journal of Anatomy*, **86**, 260.

SYLVÉN, B. (1951): On the Biology of Nucleus Pulposus. *Acta Orthopaedica Scandinavica*, **20**, 275.

IN MEMORIAM

HOLGER WERFEL SCHEUERMANN
1877-1960

Holger Werfel Scheuermann died on March 3, 1960, in Copenhagen.

Scheuermann's name is well known to the orthopaedic world. He was the first to describe juvenile kyphosis, which in many countries is now known as Scheuermann's disease.

Scheuermann was the son of a practitioner from Hørsholm, near Copenhagen. He was born on February 12, 1877, and graduated in 1902. He was trained both as an orthopaedic surgeon and as a radiologist. From 1910 to 1919 he was assistant surgeon to the Society and Home for Crippled in Denmark. From 1935 he was chief radiologist to the municipal hospital, Sundby, Copenhagen. He retired in 1947.

Scheuermann described juvenile kyphosis in 1920 to 1921, when radiology still was in its youth. His concept of the pathology—disturbances in the ossification of the end-plates of the vertebrae—is not unanimously accepted, but his description of the disease can be said to be classical and has not been improved upon by later authors.

Internationally, Scheuermann was probably the most well known Danish radiologist. Apart from his work on juvenile kyphosis his studies from 1932 on the normal and pathological sella turcica is internationally known. In 1937 he also described a radiographic technique for exposing the optic foramen, which also gives him credit. Although his work on juvenile kyphosis was original, unaccountably it was not accepted for the doctorate degree of the University of Copenhagen, and he was over eighty when the University in 1957 elected him Honorary Doctor of Medicine in recognition of his work. It was touching to witness his happiness on receiving this honour although it was given him so late.

Scheuermann had a gentle and quiet personality, and his mind was engaged in practical and scientific problems. He was always to be seen at scientific meetings not only of the Radiological Society but also of the Danish Orthopaedic and Surgical Associations. He often travelled in other countries on his scientific bent; he told me once how envious he was when visiting Schmorl because of Schmorl's immense patho-anatomical material and facilities for investigation.

We in Denmark shall always remember Holger Scheuermann for his quiet kindness and for his scientific originality in the field of orthopaedics and radiology. J. M.

PROCEEDINGS AND REPORTS OF UNIVERSITIES COLLEGES, COUNCILS AND ASSOCIATIONS

GREAT BRITAIN AND IRELAND

UNIVERSITY OF LIVERPOOL

McMURRAY MEMORIAL LECTURE

The third McMurray Memorial Lecture was given by Professor John Bruce, President of the Royal College of Surgeons of Edinburgh, in the new Medical School, Liverpool, on November 29, 1960. Professor Bryan McFarland and Mr J. Leggate, the Dean of the Medical School, presided.

Professor Bruce took as his subject the various aspects of skeletal disease as they presented to the general surgeon. He started by considering certain primary and secondary neoplasms of bone, illustrating his remarks with radiographs and pictures of tumours of the jaw and skull as well as of the limbs. He then went on to discuss the relationship between eosinophilic granuloma and Letterer-Siwe disease and the Christian-Schüller syndrome. He next showed the radiographs of a man with a hypernephroma and a secondary deposit in the scapula, who was treated by a forequarter amputation and nephrectomy and who has survived for ten years so far.

The various causes of generalised osteoporosis were considered and he pointed out that they might include the failure to form new bony matrix, the failure of the matrix to calcify, and the absorption of bone; and that in many syndromes these errors might be combined. He showed a number of striking cases of osteoporosis associated with endocrine disturbance; his display of cases of severe osteoporosis following extensive gastro-intestinal resection was both beautiful and startling, and a revelation to many orthopaedic surgeons.

Professor Bruce concluded by considering some of the problems of hyperparathyroidism and showed the radiographs of a woman with severe hyperparathyroidism whose child had suffered from hypoparathyroidism since birth. When he removed the mother's adenoma he implanted it into the child's tibia. At first there was severe hyperparathyroidism in the child, who became extremely ill and nearly died, but who ultimately survived; at the area of the implant there was considerable bone resorption, indicating that the parathyroid hormone has a localised osteolytic effect on bone. Professor Bruce suggested that there were probably two other parathyroid hormones, one acting on the kidney and the other on gastric secretion.

The lecture excited much interest and drew a large audience; both orthopaedic and general surgeons appreciated the lecture which was an admirable illustration of the very difficult problems in this fringe field to which a general surgeon with an interest in orthopaedic surgery had contributed so much.

If any of the audience had previously considered bones to be inert, Professor Bruce must certainly have convinced them that bone is very much alive and plays a vital part in the body's metabolism.

BRITISH ORTHOPAEDIC ASSOCIATION

ELECTION OF HONORARY FELLOWS, CORRESPONDING FELLOW, EMERITUS FELLOWS, FELLOWS, MEMBERS AND ASSOCIATES

At a meeting on April 14, 1961, the following were elected:

Honorary Fellows—C. F. W. Illingworth (Glasgow), Sir James Paterson Ross (London).

Corresponding Fellow—J. Neill Garber (Indianapolis).

Emeritis Fellows—W. B. Foley (Oxford), M. Forrester-Brown (Edinburgh).

Fellows—Hugh Barry (Sydney), C. J. Kaplan (Durban), Edith McComas (Melbourne), P. K. Mullaferoze (Bombay), W. A. J. Pike (Auckland), F. R. Tucker (Winnipeg).

Members—V. P. Amato (Malta), M. Anwer (Mecca), R. B. Duthie (Rochester, N.Y.), S. Y. H. Fang (Hong Kong), R. H. Freeman (R.A.M.C.), F. J. Hedden (Durban), W. C. Lawrence (Liverpool), C. R. F. McDowell (Dublin), W. A. Watt Maney (London), J. H. Miller (Glasgow), T. J. Mills (Winnipeg), T. H. Morgan (Swindon), R. Owen (North Wales), L. H. Pimm (Ipswich), E. R. Price (Barnsley), E. A. Smyth (R.A.M.C.), P. Vulfsons (Salisbury, S. Rhodesia), S. L. Weissman (Tel-Aviv), P. F. Williams (Melbourne), A. L. Zaoussis (Athens).

Associates—R. G. Burwell (Leeds), D. J. Cowan (R.A.M.C.), E. Danos (Athens), D. M. Davies (Swansea), H. J. B. Day (Manchester), H. Dovey (Liverpool), R. H. K. Gruneberg (Kirkcaldy), P. Gupta (Bristol), K. T. Hesketh (R.N.), M. Laurence (London), B. B. Mandell (London), J. G. Mander (R.A.F.), J. G. B. Myles (London), M. F. Porter (Aberdeen), D. R. Richard (London), M. P. Robinson (Oswestry), H. S. Sabbak (Baghdad), R. S. Sneath (London), T. G. Thomas (London), R. C. W. Williams (London), Ruth Wynne-Davies (Exeter).

ROBERT JONES MEDAL AND PRIZE 1961

The Robert Jones Medal and Prize for 1961 has been awarded to Mr J. A. Wilkinson of London for his essay on " Etiological Factors in Congenital Dislocation of the Hip."

SCIENTIFIC SOCIETIES AND REGIONAL ORTHOPAEDIC CLUBS

THE BONE AND TOOTH SOCIETY

Abstracts of papers delivered at a meeting held at the Royal National Orthopaedic Hospital, 234 Great Portland Street, London, W.1, on January 18, 1961.

The anatomy and pathology of the mandibular joint—*Dr H. J. J. Blackwood* (London) said that the developmental anatomy and pathology of the mandibular joint was studied in a series of sixteen human foetuses and 530 joints excised from human cadavers of varying ages. The condylar cartilage developed in the foetus at about the 40–50 millimetre crown-rump stage and the joint cavities first appeared at the 50–60 millimetre stage. Vascular canals developed in the cartilage of the 130 millimetre crown-rump foetus and persisted within the growing condylar cartilage until the third year of life. Growth of the condylar cartilage normally stopped at the beginning of the third decade.

The most common disease of the mandibular joint was osteoarthritis. This usually involved first the mandibular condyle, causing disruption of its articular surface and destruction of the intra-articular disc. Destruction of the disc appeared to cause secondary osteoarthritic changes in the articular surface of the glenoid fossa. Rheumatoid arthritis might also involve this joint and cause adhesions between the intra-articular disc and joint surfaces which could lead to complete fibrous ankylosis of the joint. Overclosure due to the edentulous state was thought to be responsible for changes in the contour of the articular surface of the mandibular condyle. Primary tumours of the joint were rare but involvement in metastatic tumour spread was less rare. The osseous components of the joint might be affected by general diseases involving the skeletal system, as, for example, in parathyroid disorders.

Electromyography of the temporo-mandibular joint—*Mr B. E. Greenfield* (London) described a standardised technique for using monopolar surface electrodes, placed bilaterally on the anterior and posterior temporalis muscles and on the lower anterior part and the upper posterior part of the masseter muscles, to record on an eight channel Grass Electro-encephalograph ink-writer. It was important that simultaneous bilateral recordings should be made. Electromyographs for various voluntary mandibular movements involving the occlusion of the teeth as well as free movements of the mandible were shown. Both the temporalis and the posterior masseter muscles were shown to be involved in closing from the wide open position to the rest position, whereas in voluntary protrusive closing the anterior masseter muscles participated and the posterior temporal muscles did not. Asymmetrical muscle activity in free movements of the mandible was frequently associated with temporo-mandibular joint disorders, and two examples were shown, as was the tracing of a patient before and after unilateral condylectomy with the eventual reversion to a normal muscle pattern for the movement of closing.

Neurophysiological aspects of joint function with particular reference to the temporo-mandibular joints—*Dr B. D. Wyke* (London) said that the coordinated contraction and relaxation of muscles that was responsible for joint movement was modulated by a combination of reflex and voluntary influences on their motor units. Both voluntary and reflex influences were closely related to afferent activity entering the neuraxis from the moving parts themselves. In the absence of an adequate proprioceptive input of this type, production of movement in higher animals was almost impossible. Recent work indicated that a considerable proportion of this input comes from receptors in and around joints. These joint receptors provide the greater part of the afferent activity upon which the perceptual awareness of joint position and joint movement depends, as well as contributing significantly to segmental and intersegmental reflex coordination of muscular activity during movement.

Four types of receptor ending had been identified in mammalian joints. Three were encapsulated end-organs; the fourth was an unmyelinated nerve network. About 60 per cent of the corpuscular endings lay in the fibrous capsule, 30 per cent in the periarticular tissues, and 10 per cent in the joint ligaments.

Type 1 was the most numerous variety, with thinly encapsulated globular endings in outer layers of the fibrous capsule, which discharged continuously at rest. Activity was increased or decreased by changes in tension or pressure in the joint capsule, and adaptation was slow. They formed receptor complexes each of which was operative over limited angles of joint movement. Activity was facilitated in some muscles, and antagonists were inhibited, and finally this type signalled the direction, rate of movement, and static position. *Type* 2 had no resting discharge and consisted of elongated conical endings in the deeper layers of the capsule, with a thicker sheath. They were stimulated briefly by rapid movements in any direction and they signalled the onset of movement and rate of acceleration, as well as vibration in joints. *Type* 3 also had no resting discharge. The fusiform encapsulated endings were in ligaments only. Stimulated by high tensions in ligaments, they adapted slowly and produced inhibition of muscular activity towards extremes of joint movement. *Type* 4 was an unmyelinated network in the deeper layers of the capsule. It was a pain receptor system and produced muscle spasm when stimulated.

Electromyographic studies on patients suggested that some receptors in the temporo-mandibular joints influenced masticatory movements, but nothing was known of the receptors—or their behaviour—in relation to these joints. Investigations had been undertaken to obtain this information, and Type 1 and Type 3 receptors had been identified histologically in the temporo-mandibular region.

Radiology of the temporo-mandibular joint—*Dr E. Samuel* (Edinburgh) said that the radiological demonstration of the temporo-mandibular joint demanded accurate technique and painstaking attention to detail. The routine projections used were lateral, oblique and postero-anterior views with or without 30 degrees angulation of the incident beam. In addition, both views were taken with the jaws closed, in the mid-position, and fully open. However, even with these views a comprehensive study of the temporo-mandibular joint could be achieved only by the use of tomographs, again with the mouth fully closed and open. Linear tomography had been used; circular tomography had not added appreciably to our knowledge. The normal anatomy of the temporo-mandibular joint shown radiographically was confined to the bony outlines of the condyle and the glenoid fossa. The width of the temporo-mandibular joint was governed by the dentition and bite as well as by changes in the joint itself. Evaluation of the width of the joint space was therefore fraught with pitfalls and arthrography had not achieved widespread use. Pathological conditions of the meniscus or joint capsule caused little change in radiographs and osteoarthritis rarely showed radiological changes; nor did rheumatoid arthritis apart from decalcification. The mandibular condyle was the seat of two special types of benign tumour—exostosis and osteochondroma. Both these tumours caused difficulties in mouth opening or caused asymmetrical opening. Deformity of the glenoid fossa was associated with exostoses. Metastatic deposits might also occur in the condylar head, sometimes with gross destruction of bone.

Surgical management of temporo-mandibular joint derangements—*Mr T. G. Ward* (East Grinstead) said that surgery on the mandibular joint was indicated only when conservative methods of treatment had failed to bring relief to the patient. There were two standard operative procedures: 1) The removal of the condylar head for osteoarthritis when degenerative changes could be demonstrated radiologically in the condylar head. These cases were rare. 2) A modification of the position of the condylar head could be effected by condylotomy. The surgical procedure was best done by the blind approach, a director being passed from behind the mandible, one finger's breadth below the attachment of the lobe of the ear, through the sigmoid notch; this was used to pull back a Gigli saw into position. The condylar neck was then sawn through, but no attempt was made to saw through the periosteum on the lateral side. The head of the condyle could be fractured forward if necessary by manual pressure. The director was passed with the mouth wide open, when it would emerge underneath the malar bone and be in the correct relation to the sigmoid notch. This operation had several advantages. First, it served to bring the condylar head forwards and inwards, and, since most painful temporo-mandibular joints were related to a posterior displacement of the condylar head, this was sound treatment. Second, it altered the proprioceptive mechanism in relation to the external pterygoid muscle on the affected side. Whether one attributed the pain of the derangement to a purely mechanical factor, to posterior displacement of the condylar head or to an upset in the proprioceptive mechanism of the muscles of mastication, the operation remained a rational procedure. It had the considerable advantage of not leaving the patient any worse off than before—a factor that should always be taken into consideration before carrying out joint surgery. Finally, the patient's stay in hospital was relatively short, three to five days being the usual period.

SOUTH-WEST ORTHOPAEDIC CLUB

The autumn meeting of the South-West Orthopaedic Club was held on November 26, 1960, at Plymouth. Mr M. Salz was in the chair.

The meeting in the morning was at the new Dame Hannah Rogers School for Cerebral Palsy at Ivybridge by invitation of the Headmistress, Miss A. A. Hill. The Chairman of the Trustees,

Mr N. Capener, gave an introductory talk. He described how the new school was financed and pointed out that, although its purpose was primarily educational, its heavy maintenance was justified by the research programme.

Treatment of cerebral palsy—*Mr G. J. Lillie* (Plymouth) described current methods of treatment at Ivybridge. The aim was to reduce spasticity and abnormal movements to improve voluntary function. Splints had been largely discarded because they increased the stretch reflex and shifted the spasticity proximally; treatment was first given to increase the stretch reflex peripherally, then, by posturing and using the tonic reflexes, to reduce spasticity in the child as a whole. After some months this produced a lasting effect and improved the existing voluntary function. Mr Lillie then described the limited use of operative treatment. Tenotomies and neurectomies were rarely used but some arthrodeses and transplants were still helpful. He stressed the need for recognition of the subluxating hip associated with coxa valga, which frequently needed surgery to prevent complete dislocation.

Early diagnosis of cerebral palsy—*Dr F. Brimblecombe* (Exeter) discussed the early diagnosis of cerebral palsy, which mostly occurred in the group of " special risk babies," such as those with premature or difficult births, rhesus abnormalities and complicated pregnancies. The two most important aids to diagnosis were delay in reaching normal milestones, and poverty of movement with persistence of primitive reflex patterns. He demonstrated these points on two babies of nine months old, one of whom was normal and the other suffering from cerebral palsy.

Demonstration—A demonstration with films and patients was then given by Mrs D. P. Beaman, Physiotherapist in Charge, on the various methods of treatment of athetoid and spastic cerebral palsies.

Cubital pain—*Mr N. Capener* (Exeter) suggested that there might be a factor called " nerve oedema " in cubital pain which might have a proximal cause, such as in the neck. This could be responsible for those peripheral conditions associated with constriction of nerves, such as the carpal tunnel syndrome and ulnar nerve neuritis. He went on to discuss " tennis elbow," which was known to be often associated with cervical spondylosis. He described cubital neuralgia due to compression of the posterior interosseous nerve. In seeking an anatomical explanation for this he had found that the posterior interosseous nerve lay beneath a deep fibrous septum and that it passed into the supinator muscle between clearly defined oblique and transverse portions where it was anatomically possible for the nerve to be constricted and thus give rise not only to the symptoms on the outer side of the elbow, but also weakness of dorsiflexion of the wrist and extension of the fingers.

Clinical demonstration—The afternoon session was held at the Mount Gold Hospital, Plymouth, where patients were shown. *Mr G. J. Lillie* and *Dr G. R. Steed* (Plymouth) showed a patient with the " stiff man " syndrome. He had gradual stiffening of all the muscles and joints of his lower limbs without significant arthritic changes. Full biochemical investigations showed that the total amount of body fluid was normal, but the extracellular fluid was six times normal, at the expense of the intracellular fluid. The patient recovered quickly on cortisone, and had since been maintained on a small dose. Investigation of pituitary function revealed hypopituitarism secondary to adrenal failure.

Mr Lillie and *Dr Steed* also showed a patient suffering from osteomalacia secondary to steatorrhoea and megaloblastic anaemia after total gastrectomy. The patient had responded to vitamin B_{12}.

Other patients were shown by *Mr Lillie* and *Mr M. Salz* (Plymouth) including cases of coeliac rickets, renal tubular acidosis giving rise to rickety bone changes, dysplasia epiphysialis multiplex, juvenile myxoedema, diastematomyelia, malignant osteoclastoma of the scapula, Brooks-Seddon pectoralis-to-biceps transplant after poliomyelitis, acute lymphocytic leukaemia with bony deposits, and a rhabdomyosarcoma in the first interosseous space of the hand of a baby of one month old, which was treated by local removal with no recurrence after two-and-three-quarter years.

Extra-articular arthrodesis of the subtalar joint for correction of paralytic flat feet in children—*Dr T. Pantazopoulos* (Athens) described the Grice operation in which the calcaneum is replaced and held beneath the talus by an extra-articular arthrodesis so as not to interfere with the growth of the foot. This removed the deforming power of the peronei and restored active inversion and dorsiflexion. The operation was indicated in the young child with increasing pes planus with lateral displacement of the calcaneum. The extra-articular arthrodesis was carried out by means of two cortico-cancellous grafts taken from the upper tibia. The grafts were so placed that they lay almost in a plane at right angles to the axis of motion of the subtalar joint and with the cancellous surfaces apposed. They were slightly oblique so that the lower ends of the graft in contact with the calcaneum were anterior to the upper edges. The pointed ends were countersunk through the cortex of the talus and calcaneum. Tendon transplants should be done when possible at the same operation. The type of transplant depended on the severity of the deformity. If there was fixed equinus deformity of such a degree that it could not be corrected at operation, the grafts were allowed to consolidate and the foot was later wedged up into dorsiflexion. In some cases posterior capsulotomy of the ankle and lengthening of the tendo

calcaneus might be necessary. The failures were due to incomplete correction or overcorrection, persistent muscle imbalance and failure of fusion. It was suggested that the procedure could be definitive and lasting and would not need to be followed by triple arthrodesis later.

Intertrochanteric displacement osteotomy using internal fixation—*Mr F. Wheeldon* (Plymouth) described his method of intertrochanteric displacement osteotomy for osteoarthritis of the hip. He had used internal fixation in all operations without immobilisation in plaster. The patients were allowed up as soon as possible and many were out of hospital within the month. After trying different types of internal fixation he now used a Capener-Neufeld nail plate. He described his results in twenty-six cases.

Approaches to lower limb vascular diseases—*Mr M. Reilly* (Plymouth) made an appeal for the immediate suturing of a cleanly divided large artery. This could be done without special instruments and was an entirely satisfactory procedure. He then described the various operations available for the treatment of arteriosclerosis, and gave the indications and contra-indications for the use of arterial grafts or prostheses, as well as for other procedures, such as sympathectomy, neurectomy and division of the calcaneal tendon.

Transcervical fractures of the femur treated by fixation with Charnley's spring-loaded screw—*Mr E. Danos* (Athens) described the results of forty-six such operations which had been done at the Winford Orthopaedic Hospital, Bristol, between April and December 1958. Thirty-seven of these were assessed, of which nineteen were successful and eighteen failed. The failures were due to avascular necrosis, non-union, and breaking out of the screw. There was a definite relationship between the results and the quality of reduction and the position of the screw. The conclusion was drawn that there was no advantage to be gained by using this appliance.

Visceral injuries complicating fractures—*Mr R. H. Mills* (Pontypridd) said that there were three types of visceral injury complicating fractures: the very severe type which caused sudden death, the more usual picture presenting as an acute abdominal emergency, and injuries with delayed manifestations, such as central rupture of the liver, silent rupture of the spleen or necrosis of the bowel following a mesenteric tear. Exact diagnosis was difficult, especially if there was a delay, and Mr Mills gave a helpful account of the ways in which diagnosis could be aided. Of special difficulty was the problem of internal injury in the unconscious patient.

The Annual Dinner was held in the evening, after which an illustrated talk on the Club's visit to Sweden was given by the Honorary Secretary, Mr R. Merryweather.

IRISH ORTHOPAEDIC CLUB

The autumn meeting of the Irish Orthopaedic Club was held in the Institute of Clinical Medicine in Belfast on November 26, 1960.

Spinal injuries—*Mr R. I. Wilson* (Belfast) said that, of over 26,000 fractures of all types dealt with in the Royal Victoria Hospital between January 1953 and June 1960, 220 or less than 1 per cent involved the spine. Of these, 13 per cent were in the cervical region, 31 per cent in the thoracic region and 56 per cent in the lumbar spine. Whereas compression fracture was the most common type seen in the thoracic and lumbar spine, 55 per cent of the cervical injuries were fracture-dislocations. Of the 220 fractures thirty-two were unstable, seventeen being in the cervical spine, nine in the thoracic spine and six in the lumbar spine. Discussing the radiological investigations Mr Wilson stressed the importance of flexion and extension lateral views in the cervical spine, and tomography and oblique views to exclude dislocation or subluxation in the thoracic spine. Open reduction and plating was done in thirteen cases; the value of internal fixation in permitting exact assessment of the cord or root damage, in preventing pain and in facilitating frequent turning and active and passive limb movements was emphasised. After operation full recovery from paralysis took place in four patients and partial recovery in two; no pressure sores developed. In the early care of the bladder a self-retaining Gibbon's catheter was used and changed every five days with washing out and aspiration, and was removed after about three weeks, by which time recovery of normal control or development of reflex emptying had been achieved.

Charnley stabilisation of the hip—*Mr B. T. Crymble* (Belfast) described his experiences in nineteen of these operations. He followed the original technique and the patients started weight bearing after six weeks in plaster. There were no deaths in the series, but two patients subsequently developed undisplaced pertrochanteric fractures. In seven patients solid bony arthrodesis was achieved, three were clinically but not radiologically solid and nine had about 20 degrees of movement in flexion and extension. The patients with solid bony union were all aged sixty or less. Eleven patients had an excellent result with no pain and a very good gait; five had slight pain on first rising from a chair,

but did not require a stick; and three were unrelieved. The members of the Club were impressed by the gait of the patients demonstrated.

Replacement of the femoral head with the Austin Moore prosthesis—*Mr John Lowry* (Belfast) reviewed a series of fifty-four cases of replacement of the femoral head by Austin Moore prosthesis; in twenty-nine this was done as primary treatment for cervical fractures, in fifteen as a salvage procedure after failure of pinning, and in ten for osteoarthritis. The average follow-up period was eighteen months and the average age of the patients was seventy-three. There were six deaths in the series. In the survivors good results with no pain and the ability to walk one or two miles were achieved in thirty-four cases, fair results with no pain and the ability to get about independently indoors in thirteen, and only one patient was classified as poor who was chair- and bed-ridden. In all, the hip joint was approached by Kocher's method and many patients were sitting out of bed within forty-eight hours. The indications for the use of this method as the primary treatment of cervical fractures was an unsatisfactory reduction, some neurological abnormality, secondary deposits and mental instability.

Facio-maxillary injuries—*Mr Roy Whitlock* (Belfast) outlined the history of this speciality and summarised the mechanism and treatment of the seven worst groups of injuries. In discussing the central block middle third fracture in which the malar bones are intact, he described the construction of silver cap splints and stressed that bony reduction must precede soft-tissue repair. In the high transverse (transorbital) fracture, fixation was obtained by means of a Steinmann pin passed right through the face and fixed to supporting rods and plaster-of-Paris. When malar and maxillary fractures were combined the malar displacement should be dealt with first.

After lunch in the Royal Victoria Hospital the Club visited the various departments of the Institute of Clinical Medicine, including the Departments of Pathology and Microbiology.

SOUTH AFRICA

SOUTH AFRICAN ORTHOPAEDIC ASSOCIATION

The Annual Congress of the South African Orthopaedic Association was held at Cape Town from October 13 to 15, 1960. The President, Mr R. C. J. Hill, was in the chair. The meeting was preceded by an instructional course at the Karl Bremer Hospital, organised under the aegis of the College of Physicians, Surgeons and Gynaecologists of South Africa, and was the first of its kind in that country. Distinguished guests from overseas included Dr Leo Mayer (New York), Mr F. W. Holdsworth (Sheffield), President-elect of the British Orthopaedic Association, Mr R. G. Pulvertaft (Derby), Cripple Care Lecturer for 1960, Dr Beckett Howorth (Connecticut), Dr Paul S. Harmon (San Francisco), and Mr E. A. Nicoll (Mansfield). Dr Leo Mayer and Mr R. G. Pulvertaft were elected members of the South African Orthopaedic Association.

Functional anatomy of the pelvic joints—*Mr G. F. Dommisse* (Pretoria) showed that the symphysial ligaments in the adult male had a breaking strength in excess of 520 pounds. Using mathematical calculations he had found that the symphysis was subjected to tension rather than compression and that this tension amounted to a force of 600 pounds with each stride of the athlete at full speed on the level. Symphysial separation in the pregnant female normally averaged about 1 centimetre. This was controlled by an oestrogenic hormone and was reversed during the puerperium by an unidentified hormone. Reversal might be delayed, defective or inhibited, resulting in symphysial and sacro-iliac instability. Post-partum symphysial separation was a well defined clinical syndrome. A similar syndrome had been found to follow miscarriages, hysterectomy, ovariectomy and the menopause, the latter sometimes associated with osteoporosis. Mr Dommisse went on to say that the sacro-iliac joints almost always showed degeneration after the third decade, and that his experiments with macerated pelves indicated that radiographic changes at the sacro-iliac joints were difficult if not impossible to detect, but changes at the symphysis could be detected whenever present. The lumbo-sacral trunk (fourth and fifth lumbar roots) had a very close anatomical relationship to the sacro-iliac joint, and might be partly or completely severed in fractures of this region, or irritated by other lesions such as infections, osteoarthritis or post-partum instability of the pelvis, to give sciatica. An operation for the exposure of the lumbo-sacral trunk had been evolved.

Diagnostic localisation in lumbar disc disease—*Dr Paul S. Harmon* (San Francisco) emphasised that the clinical localisation of a deranged lumbar disc was of the first importance. He used routine radiography including flexion and extension films and showed that myelographs might be misinterpreted and that normal or equivocal results in the presence of abnormal discs were common. In 300 cases

he had injected physiological saline under visual control into the lumbar discs during the performance of the anterior operation. If more than 1 millilitre could be injected it was significant of disease, and it was the most sensitive and reliable method of detecting degenerated discs in the lumbar area. He had come to this conclusion after comparing the saline injections with myelography in 150 of the same cases.

Intervertebral disc excision and lumbar spine fusion by a transperitoneal abdominal approach—*Mr S. Sacks* (Johannesburg) in a report on eighty cases said that the main indication for this procedure was prolonged backache, with or without pain in the legs due to old-standing disc degeneration, with failure to respond to conservative treatment, and many other conditions such as spondylolisthesis, Pott's disease, failed fusions and backache after fractures. All the operations had been done in the past two years. The results were so far satisfactory, and on clinical assessment 88 per cent of the patients were symptom-free or improved. Forty patients had been reviewed less than six months after operation. *Professor C. E. L. Allen* (Cape Town) mentioned that in this approach the posterior structures responsible for the symptoms and signs were not visualised. *Mr Hamilton Bell* (Cape Town) expressed surprise at the number of cases operated on in such a short period. *Dr Paul S. Harmon* (San Francisco) stated that he had performed this operation on 450 cases and he found that the transperitoneal route was easier initially, but he later employed the retroperitoneal approach.

Fractures of the spine and their treatment—*Mr F. W. Holdsworth* (Sheffield) classified spinal fractures into stable and unstable varieties. The treatment of the stable variety was little or none beyond bed-rest until the acute pain subsided. The unstable varieties included fracture-dislocation and the " slice " fracture with overriding of the posterior facets, which was associated with extensive posterior soft-tissue damage, and which required stabilisation. *Mr Hamilton Bell* (Cape Town) asked what the degree of recovery was after plating for unstable spinal fractures associated with paraplegia. Mr Holdsworth replied that the plate fixation was intended to render the nursing of these patients easier. *Mr G. T. du Toit* (Johannesburg) asked if the bodies in the stable type of wedge-fractures collapsed again after reduction and immobilisation for three months in hyperextension plasters. Mr Holdsworth replied that a satisfactory plaster in extreme hyperextension could not be applied, and the results were worse than if the patients were left untreated. *Mr G. Bickerton* (Durban) asked for how long patients with stable fractures were kept in bed, and whether they were given physiotherapy. Mr Holdsworth replied that these patients were allowed to get up when they felt like it—usually about three weeks after injury—and that little physiotherapy was given.

Origin of congenital anomalies and the control of bone form—*Mr T. L. Sarkin* (Cape Town) presented experimental and clinical evidence to show that the present theories of bone growth as originally set forth by John Hunter were only partly correct. Using a series of ingenious models and slides, he explained his views on why the greatest growth in the length of the lower limbs was from the distal end of the femur and proximal end of the tibia, whereas the greatest growth in length of the upper limb was from the proximal end of the humerus and the distal ends of the radius and ulna. On the basis of his theory of helical growth he proposed new explanations for various growth anomalies.

An experiment in rehabilitation—*Dr I. Guthrie* (Somerset West) described the establishment of a rehabilitation centre for injured and ill workmen at the African Explosives and Chemical Industries factory. This centre was the first started by a private industrial concern in South Africa. The initial inspiration was provided by the Vauxhall Rehabilitation Film. Unlike the Vauxhall Motor Works there was a comparatively small number of employees and a lack of repetitive production jobs at the factory. He had the same difficulties to face in regard to the attitude of the management and the trade unions.

The experimental use of a muscle graft in the regeneration of a peripheral nerve—*Professor J. Kirsten* (Stellenbosch), after discussing the standard methods of nerve suturing and grafting, said that there were certain disadvantages to nerve grafting and much experimental work had been done to find a substitute for the nerve graft, including the use of blood vessels, synovial sheaths, plastic tubes and allantoic membrane in an endeavour to construct a suitable tunnel along which to guide the regenerating axons into the distal Schwann sheaths, but without success. In his Hunterian Lecture in 1955 on the Functional Importance of the Blood Supply of Peripheral Nerves, Professor G. Causey had observed that nerve axons regenerate through a segment of muscle interposed between two cut nerve ends, and that it would be highly advantageous if one could get satisfactory regeneration of a nerve through such a muscle graft, since much muscle was available for use as grafting material. Professor Causey had performed a number of experiments in this direction and had subsequently introduced this subject to Professor Kirsten. Although the work was still in progress some results were worth mentioning. The nerve to the medial head of the gastrocnemius in the rabbit was isolated and one or two centimetres were removed. A piece of muscle one centimetre thick was excised from the biceps femoris and the

proximal and distal ends of the severed nerve were buried in the muscle graft. On one side a completely avascular muscle graft was used and on the opposite side the graft retained an intact arterial supply. The nerve and the muscle graft were isolated at varying intervals from ten to forty weeks after the operation and the nerve was stimulated with electrodes both proximal and distal to the graft. The isolation of the nerve was sometimes difficult and often impossible, but when it could be done there was good contraction of the gastrocnemius as well as the graft on proximal stimulation of the nerve, and good contraction of the gastrocnemius on distal stimulation. Sections of the nerve proximal and distal to the graft were then removed and fixed in osmic acid. The nerve sections were prepared according to a modified Weigert-Pal method and examined microscopically. On the whole it appeared that the avascular graft was as good as, if not better than, the vascular graft.

Recurrent dislocation of the shoulder—*Mr T. B. McMurray* (Cape Town) said that it was generally held that the question of recurrent dislocation of the shoulder was settled at the time of the initial injury. He contended that there were two additional factors that might increase the likelihood of recurrence. The first was Kocher's method of forced lateral rotation, which could extend an initial, but trivial, capsular or labral tear. Of sixty-four dislocations of the shoulder reduced by Kocher's method, twenty-seven (40 per cent) became recurrent; but of 112 dislocations reduced by lifting the head of the humerus into place, thirteen (12 per cent) became recurrent. The second factor was the after-care of dislocations. The early exercises and physiotherapy which had been used after the first dislocation in 70 per cent of recurrent cases may have militated against the healing of the torn tissues at the front of the joint. Mr McMurray briefly discussed the disadvantages of the Bankart and Putti-Platt operations, and described his procedure which was a modification of Bristow's operation. The distal half-inch of the coracoid process with its three muscular attachments was transplanted through a transverse slit in the subscapularis muscle into the rawed glenoid rim and fixed with a single screw. The only two recurrences out of seventy-three operations performed since 1948 were caused by unusual and severe injuries.

Hazards in orthopaedic surgery—*Dr I. S. de Wet* (Pretoria) gave first the history of a European man aged thirty-nine years who, twenty-one years previously, had had a septic arthritis of the left hip. Five years previously a McMurray osteotomy—with minimal displacement—had been performed for pain and stiffness. He had some relief and was able to continue farming until 1960 when more pain forced him to seek advice again. Clinically the hip had only 30 degrees of flexion and investigations revealed no signs of active disease. A Charnley arthrodesis was performed but during the operation while dense fibrous tissue was being separated from the edges of the acetabulum, the surgeon's finger penetrated the external iliac artery. Through an abdominal approach it was repaired with a seven-inch woven Teflon graft. The entire procedure lasted seven and a half hours and eleven pints of blood were given to the patient. The patient survived after a stormy convalescence and at four months there was bony ankylosis of the left hip and both anterior and posterior tibial arteries were pulsating.

Another hazard was in a woman aged forty-two who presented with intermittent pain and stiffness in her neck. Later she developed numbness of the whole of the right arm and weakness of grip. She had limitation of cervical and spinal movements in all directions, with wasting of the intrinsic muscles of the hand, especially those of the thenar eminence, and hypoaesthesia. Radiographs showed narrowing of the disc spaces between the fifth, sixth and seventh cervical vertebrae. After conservative treatment a manipulation under a general anaesthetic produced some relief. Later, after myelographic and angiographic examinations, syringomyelia was diagnosed. Five months later, after developing spastic quadriplegia, the patient died. Necropsy revealed a meningioma about three centimetres in diameter in the posterior fossa of the skull close to the foramen magnum.

As his third example Mr de Wet told of an eighty-three-year-old lady who presented with an acute exacerbation of long-standing osteoarthritis of the left knee. The knee was aspirated and 1 millilitre of Codel-cortone was injected with prompt relief of pain. A week later she returned for an injection into the opposite knee because of pain. This was done and three days later the patient developed a fulminating septic arthritis of the knee and died of a staphylococcal septicaemia forty-eight hours later.

Scalenotomy and its place in the cervico-brachial group of symptoms—*Mr L. Mirken* (Port Elizabeth) reviewed fifty cases of scalenotomy performed in the last ten years. He considered the most important positive sign in the diagnosis to be tenderness over the insertion of the scalenus anterior and the reproduction of symptoms by digital pressure over the insertion at the first rib. He performed a complete section of the scalenus anterior over the subclavian artery and stripped the artery at the level of the arch.

Treatment of flexor tendon injuries in the hand—*Mr R. G. Pulvertaft* (Derby) said that our concern today was to establish a plan of treatment that could be expected to offer a good general standard of

result. He pointed out the difference in approach to the repair of a single tendon injury in an otherwise normal hand and the tendon division in a severely damaged hand. In the former restoration to normal, or near normal, should be achieved in about 75 per cent of cases, but in the latter, although improvement might be slight, it was of value. The principles and techniques of repair in the different areas of the hand were discussed. Primary repair of flexor tendons when wound conditions permitted was advisable for profundus divisions distal to the sublimis insertion, for flexor pollicis longus divisions in the distal part of the thumb, and for divisions in the palm and at the wrist level. Skin suture only and secondary repair by tendon grafting was recommended for tendon divisions between the distal palmar crease and the insertion of sublimis and for divisions of flexor pollicis longus in the palm and the base of the thumb. The operation of tendon grafting to restore profundus in the presence of an intact sublimis was described and illustrated by a film. The advantages of using plantaris as a graft were mentioned, as was the bridge graft technique for closing gaps between tendon ends in the palm and above the wrist. The problems of injuries occurring in infancy and the time lag between injury and elective repair were discussed, and several cases were shown with satisfactory results even when the delay had been over ten years. The importance of testing the amplitude of movement of the original muscles in these cases was pointed out, and the advice given that sublimis of another finger should be used if the original motor appeared to be ineffective. In conclusion, Mr Pulvertaft said that the treatment of these injuries would always present a difficult technical problem. *Dr Leo Mayer* (New York) thanked Mr Pulvertaft for his masterly presentation and excellent results. *Mr J. J. Commerell* (Cape Town) asked whether early or later movement after tendon grafting was advisable for the average operator. Mr Pulvertaft replied that late movement was safer and as satisfactory. *Mr C. Möller* (Johannesburg) wanted to know what was considered the least number of pulleys advisable. Mr Pulvertaft replied that one was necessary—over the middle of the proximal phalanx. *Mr G. T. du Toit* (Johannesburg) asked whether Mr Pulvertaft was in favour of post-operative cortisone administration. Mr Pulvertaft replied that he was not in favour of this because of the danger of interference with healing. In reply to Dr Leo Mayer, Mr Pulvertaft said that he sutured digital nerves at the time of primary treatment.

Pelvic obliquity in poliomyelitis, its recognition and treatment—*Dr Leo Mayer* (New York) emphasised that he was dealing only with two types of fixed paralytic pelvic obliquity. In the first type, paralysis of the hip adductor muscles allowed the abductor muscles to produce a fixed abduction deformity which, in time, could create pelvic obliquity of two to four inches. At operation he cut the gluteal and pyriformis attachments to the tip of the greater trochanter; also he excised a portion of the capsule of the hip to allow complete correction. He then transposed a long strip of fascia lata—dissected upwards from the distal half of the thigh—to the inguinal ligament. The tension of the fascia must be sufficient to maintain the limb in a neutral position. The second type of pelvic obliquity was caused by unilateral paralysis of the external and internal oblique muscles of the abdomen, almost always associated with paralysis of the quadratus lumborum. This could be corrected by the Roger-Anderson push-pull apparatus. Sometimes tight bands of fascia or muscle had to be divided before correction could be secured. After the deformity had been corrected the fascia lata was transplanted to join the ninth rib to the iliac crest on the paralysed side. Dr Mayer added that Clark of Great Britain and Axer of Israel left the tensor fasciae muscle attached to the fascia lata when transposing it, in the hope of achieving active correction; he had used this method in his last five cases and secured even better results than with the free transfer.

Mid-carpal instability—*Mr A. J. Helfet* (Cape Town) reviewed the anatomy of the mid-carpal joint in which not more than one-third of flexion-extension took place, the rest of the movement being in the radio-carpal joint. In disorders of the mid-carpus the limitation of movement was always less than one-third of the normal range, and a painless arc of movement was always present, with pain at the extremes. Tenderness was felt in the anatomical snuff box and on the dorsum of the wrist over the mid-carpal joint. In the first stage of instability there were no radiographic changes and treatment was by manipulation under anaesthesia. In the second stage erosion of the mid-carpal joint might show on the radiograph. Mid-carpal arthrodesis was successful in relieving symptoms and restoring painless function. In the third stage the same clinical disabilities were associated with an ununited fracture of the scaphoid. Mid-carpal arthrodesis was indicated in treatment; and he had performed twenty-eight such arthrodeses. *Mr S. Sacks* (Johannesburg) pointed out that bilateral os centrale has been found to be a cause of painful wrists.

University of Cape Town artificial limb—*Professor C. E. Lewer Allen* (Cape Town) described and demonstrated the new limb and discussed its principles, and the advantages at the hip, knee and ankle joint levels. Only one limb was in full use so far. The limb had been fully described in a previous paper. *Mr H. Bell* (Cape Town) asked how the weight of the new limb compared with that of the

standard limb. Professor Allen replied that it was just over seven pounds (the standard limb weighed seven pounds).

The internal fixation of fractures—is it worthwhile?—*Mr R. C. J. Hill* (Durban). This paper will be published in full at a later date.

Double oblique telescoping osteotomy—*Mr J. J. Commerell* (Cape Town) showed a film of his technique of correcting deformity using a double oblique telescoping osteotomy.

The fibular by-pass operation for non-union of the tibia—*Mr H. Bell* (Cape Town) had first used this technique in children with defects in the tibia due to osteomyelitis and sequestration and when the standard grafting techniques had failed. He was so impressed with the results in these patients that he had extended the use of the operation to any case of fracture of the tibia with loss of bone or skin or infection which would prevent grafting, or of failed tibial graft operations. He had found it important to perform the osteotomy just below the neck of the fibula, and obliquely downwards and inwards so that a large raw surface was opposite the tibia. The fibula was held in position with one screw; an osteotomy of the fibula lower down was necessary when there was firm fibrous union of the tibia.

Anterior cervical spinal fusion—*Dr Paul S. Harmon* (San Francisco) showed a film of his operation for anterior cervical fusion.

Tibialis posterior tendon transfer—*Mr A. Singer* and *Mr M. Singer* (Cape Town) demonstrated six children in whom this operation had been performed for congenital club feet which had relapsed after conventional treatment. The children shown were from a series of thirty-six on whom this operation had been done. Three adults were shown in whom the operation had been successfully performed for traumatic lateral popliteal nerve paralysis, and the results of the operation in eight patients with paralysis after poliomyelitis were discussed.

Perthes' disease—*Mr A. Singer* and *Mr M. Singer* reviewed sixteen patients with this condition in twenty-four hips. Four unusual modes of onset were: unilateral signs with bilateral radiographic changes; a unilateral limp only, with bilateral radiographic changes; pain in the left iliac fossa and intermittent limp; and slight limitation of movement and no radiographic changes for two months. With four different methods of conservative treatment the results were the same, but the prognosis was better the nearer the onset was to the age of three. They maintained that the fate of the head was predetermined.

Rotation-varus osteotomy—*Mr A. Singer* and *Mr M. Singer* showed two examples of this operation, one being done after the successful reduction on a frame of a unilateral congenital dislocation of the hip in a child untreated until the age of four years. The other was done in a patient who had had paralytic dislocation of the hip after poliomyelitis.

Rehabilitative surgery in cerebral palsy—*Mr T. L. Sarkin* (Cape Town) said that spasticity was restricted to certain muscles and was not generalised. He showed a patient in whom he had excised the sublimis tendons with considerable improvement in the function of the hand.

A method of localised fusion of the wrist joint—*Mr S. Schwartz* (Cape Town) discussed the principles of a new method of localised fusion between the distal end of radius and the scaphoid. This fusion had all the advantages of an arthrodesis of the wrist and none of the disadvantages, because it preserved the inferior radio-ulnar joint and triangular fibrocartilage so that pronation and supination were unimpaired, and because there was no interference with the intercarpal and carpo-metacarpal joints. The column of bone bridging the radio-scaphoid joint had been found to be adequate even in those doing strenuous physical work. The indications were for a flail wrist or for injuries of the wrist joint, and especially ununited fracture of the scaphoid. He described the technique of the operation in which he used a Forstner auger bit to make a cylindrical cavity in the radius and scaphoid which was filled with autogenous cancellous bone. He had performed the operation in sixteen patients and bony fusion had occurred in all instances in about three months.

Aorto-iliac occlusive disease—*Professor J. H. Louw* (Cape Town) described the clinical findings, pathology and treatment in sixty-seven consecutive cases of aorto-iliac occlusive disease. He said the cases were divisible into those in which a localised segment was involved with the rest of the cardio-vascular system being relatively normal, and those in which there was a diffuse disease. Claudication, not only in the calf, back and thigh, but even in the hypogastrium, had led to errors of diagnosis. The claudication was, however, quantitatively and qualitatively related to effort in the muscle group affected, and was not always severe but might be felt as a dull ache, cramp, burning or discomfort. The disease, usual in the sixth decade, could occur in younger people. Signs of severe ischaemia were uncommon. The femoral pulses were palpable in two-thirds of the limbs, and the popliteal and

pedal pulses in about one-third, even with complete aortic and iliac occlusion; but the pulses were usually weaker than normal. It was most important to bear the condition in mind in every person complaining of low back pain, either alone or in conjunction with pain in any part of the lower limb. Oscillometry would assist in diagnosis, but since aortography was not without risk it was used only before operation or in certain cases of special difficulty. Treatment consisted mainly in lifelong supervision, with low-lipid diet, abstinence from smoking, care of the feet, and graduated exercises as the mainstays. Anticoagulant therapy was frequently used in conservative treatment and surgery was only an incident in the overall management. The surgical procedures used in this series were endarterectomy in fifty-six patients, endarterectomy and a Teflon by-pass graft in six, a by-pass graft alone in three, and an extensive lumbar sympathectomy in two. Success was obtained in 83 per cent with a mortality of 1·5 per cent.

DENMARK

DANISH ORTHOPAEDIC ASSOCIATION

The Association held two meetings during 1960. At the meeting held in Copenhagen in May the following subjects were discussed.

Striated muscle tissue—*Dr F. Bro-Rasmussen* and *Dr O. M. Hansen* (Copenhagen) gave a short review of the histology and development of striated muscle tissue. The morphology and topography of the terminal branches of the nerves and the motor end-plates were emphasised in relation to the increased diagnostic possibilities from the use of modern tissue stainings.

Arthrodesis of the hip with the help of a bone flap—*Dr K. Jansen* (Copenhagen) reviewed the clinical and experimental reports on the use of bone flaps. He had operated on thirteen hips using a flap of bone from the trochanter, after removing the cartilage from the joint. Internal fixation was used in most cases. The patients were between thirty-one and sixty-four years of age, and sound bony fusion was the result in all cases. He found the method especially useful after the removal of a Judet prosthesis.

Carpal tunnel syndrome—*Dr J. Rasmussen* (Copenhagen) described the symptoms in twelve patients operated on for carpal tunnel syndrome. One of the patients had also a trigger finger. At operation the carpal ligament was incised but the tunnel was reformed using a bridge made from the palmaris longus tendon. Ten patients were followed for three to eighteen months, and results were good in all but one, who had cervical osteoarthritic changes in the cervical spine. The atrophy persisted in the thenar muscles in three hands.

Severe slipping of the upper femoral epiphysis—*Dr St Adorjan* (Copenhagen) gave an account of eighteen wedge osteotomies performed for severe slipping of the upper femoral epiphysis which were done over a six-year period. The patients were followed, on an average, for four years; six had very good results, three had bad results, and five had good and four fair results. No complete necrosis was observed in this series, but three patients with partial necroses recovered and obtained fair results. Among those with bad results one developed a pseudarthrosis, and two had cartilage necrosis, which was already present before the operation. The latter should not be operated upon. From a study of the literature Dr Adorjan was able to emphasise that avascular necros is of the femoral head and of the joint cartilage might occur spontaneously and was not always a result of manipulation or operation. Since no other treatment offered better results in the good group than wedge osteotomy, it was felt that this method of treatment should not be abandoned. The same method of treatment was used in the other two departments of the Orthopaedic Hospital in Copenhagen.

The subject of the second meeting of the Association in Copenhagen in October was congenital malformations.

Embryological development with special reference to congenital deformity—*Dr F. Bro-Rasmussen* (Copenhagen) said that the zygote was formed by fertilisation of the ovum, and from the fertilised zygote there were developed the specialised tissues and organs by mitotic division, cell migration, and differentiation. This process depended on a continuous pattern of direct or indirect chemical reactions, depending on the chromosome. It seemed that enzymes were formed which worked only when surrounding conditions were suitable; and these conditions were dependent on the embryo's chemical milieu, which was itself dependent on the mother organism. For this reason the same congenital deformity could result either from genetic defects in the enzyme or enzyme system, or from an unfavourable environment created by exogenous influences from the mother organism at the time of

action of the enzyme system. There was a precise embryological age for the formation and maturity of different tissues and organs, when most cells were in a state of mitotic division and differentiation; it was during these periods that tissues were most susceptible, and the type of deformity caused by exogenous influences would therefore depend on the phase the embryo was in when it was exposed to them. Since the same deformity could arise from a variety of exogenous influences, this had been interpreted as a non-specific interference with the embryo's metabolism. Recent investigations showed, however, that exogenous factors might attract specific chemical processes in the embryo, varying with the factor involved. That exogenous influences could modify the precise structure of the gene pattern in the embryo strongly suggested that congenital deformities could result from a combination of genetic and exogenous factors.

The influence of genetic factors on congenital deformity—*Dr J. Schultz-Larsen* (Copenhagen) said that there were many things that pointed to the influence of hereditary factors on congenital deformity: there were clear dominant or recessive familial traits; there was increased incidence in affected families; and there was increased incidence in the children of consanguineous parents. On the other hand, the frequent anomalies found in deformities in uniovular twins showed that the influence of environment was important. In abnormalities due to biochemical defects the genealogical data suggested an inherited abnormality which followed the Mendelian laws, and which could be traced back to a single pair of genes. When there was a deformity of a single organ the position was more complicated, however, because that organ formation was dependent on a complex of gene pairs, which in rare combinations could give rise to congenital deformities arising *de novo*; and since deformities arising from genetic causes and from exogenous influences could be identical it might be impossible to determine which was at fault. In the last one and a half years many examples of aberrant chromosome counts and morphology had shown the importance of genetic factors in complicated congenital deformities, but there would doubtless remain a majority of genetically conditioned abnormalities in which the abnormal karyotypes would not be found even with the finest techniques of chromosome morphology; for example, when an unfortunate recombination of genes resulted in a deformity without any change in the surrounding milieu.

The influence of exogenic factors on congenital malformations—*Dr O. M. Hansen* (Copenhagen) surveyed the literature of experimental teratology and stressed the non-specificity of the influence of the different exogenic factors on congenital malformations. The resultant malformations were related to the time the factor was working during embryogenesis. In clinical work the retrospective method gave little waste in material, but was inexact. The prospective method was exact but wasteful of material. Congenital malformations must be considered to be the result of an interaction of genetic and exogenic factors.

The inherent risk of ionising radiation with special reference to orthopaedic radiography—*Dr E. Hammer-Jacobsen* (Copenhagen) said that animal experiments showed that there was no minimal threshold for mutations caused by ionisating radiation. Moreover there was a direct relation between dose and number of mutations; the effect was cumulative and independent of the quality of the x-rays, but increased with intensity. During the first two to six weeks the foetus was especially vulnerable to congenital deformity induced by radiation. The dose-effect curve varied in different animals, probably because of different thresholds. More experiments were needed, particularly in the dose range from one to ten röntgen; few reports had been published of the condition in the human. He went on to say that if one million people received a bone marrow dose of one röntgen it would probably result in one case of radiation-induced leukaemia each succeeding year; and that it was most important that those working with x-rays should understand the risks involved. Radiology departments should, therefore, employ specially trained nurses, so that they should have a full knowledge of x-ray protection, for which the most important rules were mentioned.

The teratomatous effect of vitamin A deficiency in pigs—*Mr Birthe Paludan* (Copenhagen) gave a preliminary report of his work in the last three years at the Royal Veterinaerog Landbohøjskoles Department for Physiology and Endocrinology, in collaboration with the Department of Pathology and Morbid Anatomy and the State Vitamin Laboratory. Experimental investigations had been conducted into the utilisation of oral and parenteral vitamin A in pigs, of the transfer of the vitamin from sow to offspring, and the teratomatous effect of vitamin A deficiency. Nine experimental cows were starved of vitamin A, some during the whole pregnancy, others during the first third, which was confirmed by liver biopsy and other tests. The sows gave birth to ninety-one piglets, all of which showed deformities. In all there were varying degrees of micro-ophthalmia, and included among the other common deformities were heart defects, diaphragmatic hernia, absent, displaced and deformed kidneys, and hypo-genitalia with internal hydrocephalus and spinal cord protrusion.

Irradiation-induced deformities—*Dr L. Jacobsen* (Copenhagen) said that in view of the danger of irradiation-induced deformities, and in order to investigate their prophylaxis, a register of children of all women exposed to x-rays in the first four months of pregnancy had been started in June 1959 at the Arvebiologisk Institute. The purpose was to find out if exposure to x-rays induced embryological deformities, and when this large series had been analysed the Institute intended to follow up the children. Mice *in utero* had been irradiated with 100 röntgen and the deformities that resulted and that were statistically significant were a variation in tail length, a diminution and disappearance of the ossification centres in cervical, thoracic and lumbar vertebrae, and splitting, shrinkage and deformity of the sternum. Also the irradiated animals in this preliminary investigation tended to have a smaller size and weight of litters, as well as a diminution in the mother's weight during pregnancy.

HOLLAND

THESES FOR DOCTORATE DEGREE

The theses upon which the following four abstracts are based were recently accepted for Doctorate Degree of the University of Utrecht.

Fractures of the proximal end of the radius—*Dr P. van Rosevelt* (Amsterdam) had studied 1,095 fractures of the head and neck of the radius. After a short anatomical and historical account of the elbow joint he classified the various types of injury to the upper end of the radius. It was usually caused by a fall on to the outstretched hand, and was more common in women than men. In the child the epiphysis was usually involved. From the results of 1,095 injuries it was found that simple longitudinal fractures and undisplaced marginal fracture of less than half the radial head were best treated conservatively and equally well in plaster or sling, though the latter appeared to restore function quickest. Surgical treatment was necessary for displaced fractures whether marginal or comminuted. Conservative methods were indicated in fractures of the neck and an angulation of 45 degrees was acceptable in young children, but only 15 degrees in older children and adults. Dr van Rosevelt warned against excising the radical head in children. The complications of this injury included myositis ossificans, particularly after surgery, and radio-ulnar synostosis, particularly in children.

A study of the numbers of spastic children needing treatment—*Dr J. van der Meer* (Amsterdam) found, in a survey of a rural district, sixty-nine spastic children out of a total population of 125,000. Thirty-two children were suitable for treatment. From these figures the requirements of the country as a whole were assessed. Out of 170 spastic children studied since the disease had been diagnosed there were prenatal causes in fourteen children, including rubella, parotitis, eclampsia and the anti-Rhesus factor; in thirteen there had been difficulty in parturition; two were delivered by Caesarian section, forty-one were premature, forty suffered from severe asphyxia or other immediate post-natal complications; twenty-eight had later post-natal complications such as cerebral infections or trauma; and in thirty-two there was no information. Twelve of the children were twins. Usually the disease was diagnosed at about six months of age. Treatment started early gave the best results, but the nation-wide facilities for treatment could be improved.

Experimental muscular hypertrophy in the rat—*Dr B. van Linge* (Groningen) described an experimental method of causing the plantaris muscle of the rat to hypertrophy. The muscle was sutured to the calcaneum, and the gastrocnemius totally denervated to allow the plantaris—which is one-fifth the size—to do all the work. The rats were then exercised for up to nine hours a day. The plantaris could hypertrophy to twice its normal weight; histologically there was also a true hyperplasia with the formation of new muscle fibres, and there was an increase in myoglobin which accounted for the darker colour of the muscle. After prolonged exercise some sporadic hyaline degeneration was seen. Dr van Linge emphasised that if a muscle was surrounded by a tense fascia, pressure necrosis might occur with hypertrophy, such as in the extensor hallucis longus muscle in man.

Treatment of anaerobic infections with oxygen under pressure—*Dr I. Boerema* and *Dr W. H. Brummelkamp* (Amsterdam) said that the treatment of anaerobic infections by the inhalation of oxygen under a pressure of three atmospheres had been studied in the Wilhelmina Gasthuis surgical clinic at the University of Amsterdam. Three critically ill patients—two of them had Clostridium Welchii infections with gas gangrene—recovered with this treatment.

ANNOUNCEMENTS

BRITISH ORTHOPAEDIC ASSOCIATION AUTUMN MEETING, 1961

The autumn meeting of the British Orthopaedic Association will be held in London from September 28 to 30, 1961. Members of the Italian Orthopaedic Society have been invited to attend the meeting. The morning of the first day, September 28, will be devoted to an instructional course, open upon payment of the entrance fee not only to Fellows, Members and Associates of the British Orthopaedic Association but to all those interested. Those wishing to present papers at the meeting should submit an abstract of approximately 400 words to the Honorary Secretary of the British Orthopaedic Association, 47 Lincoln's Inn Fields, London, W.C.2. As already announced, the closing date for receipt of abstracts is May 31.

UNIVERSITY OF OXFORD

NUFFIELD ORTHOPAEDIC CENTRE

The following is the Lecture Programme for Trinity Term, 1961, at the Wingfield-Morris Orthopaedic Hospital, at 8.30 in the evening. May 4—"Growth and Nutrition from the Paediatrician's Viewpoint," Dr V. Smallpeice; May 18—"The Healing of Fractures," Professor P. Lacroix; and June 1—"Recent Advances in the Understanding of Osteogenesis," Professor J. Trueta.

HOLLAND

CONFERENCE ON THROMBO-EMBOLISM IN SURGERY

A conference on the prevention of thrombo-embolism in surgery has been arranged at the Dijkzigt Hospital, Rotterdam, on May 27, 1961. Those wishing to attend should write to the Secretary, Dr J. M. Greep, Surgical Department, Dijkzigt Hospital, Rotterdam, Holland.

Book Reviews

Osteochondritis Dissecans. Loose Bodies in Joints; Etiology, Pathology, Treatment. By I. S. SMILLIE, *O.B.E.*, Ch.M., F.R.C.S.(Ed.), F.R.F.P.S., Lecturer-in-Charge, Department of Orthopaedic Surgery, University of St Andrews; Adviser in Orthopaedics, Eastern Region (Scotland) Hospital Board; Surgeon-in-Charge, Fracture and Orthopaedic Department, Royal Infirmary, Dundee, and Fracture and Orthopaedic Unit, Bridge of Earn Hospital, Perthshire. 10×7 in. Pp. viii+224, with 231 figures, many in colour, and 7 tables. Index. 1960. Edinburgh and London: E. & S. Livingstone Ltd. Price 60s.

This monograph is divided into two main parts of almost equal size. The first part relates to the etiology and pathology of the condition and is divided into a general review of the theories of causation and pattern of the disease on the one hand, and a consideration of individual joints on the other. The second part deals with the author's extensive experience of the condition as seen in the knee, elbow, ankle and hip joints, and includes an account of Freiberg's infraction of the head of a metatarsal.

Mr Smillie divides his cases broadly into two groups, juvenile and adult osteochondritis dissecans. He believes that anomalies in ossification can be observed up to the age of ten, and that these may determine the clinical onset of osteochondritis dissecans in the second decade. It is suggested that this is the normal pattern of development in the juvenile cases, but there are two qualifications. The first is that anomalies of ossification do not necessarily lead to the development of osteochondritis dissecans. The second is that some cases observed in the second decade of life may fit into the adult pattern rather than the juvenile. Mr Smillie claims it is possible to trace transition from the observed anomaly of ossification into established juvenile osteochondritis dissecans. Trauma is thought to be the factor which localises and determines the development of osteochondritis dissecans in this group. The local manifestation is commonly part of a constitutional abnormality which Mr Smillie calls "enchondral dysostosis."

In the adult group of cases trauma is thought to be the main or perhaps the sole factor in determining the onset of osteochondritis dissecans. The trauma may be endogenous and/or exogenous.

The author also describes tangential osteochondral fractures, but only to exclude these from true osteochondritis dissecans.

The second half of the book dealing with case reports and treatment includes a series of 165 patients in whom the knee joint was affected. Of these, thirty patients were treated by drilling, and thirty by drilling and internal fixation using a special metal pin which the author has devised for this form of treatment of which he has been the pioneer.

The illustrations and radiographs are excellent and the publishers have again lived up to their high reputation in the standard of production. The book will undoubtedly be a standard work of reference on the subject, and the author is to be congratulated.—Norman ROBERTS.

Verletzungen bei Kindern und Jugendlichen. By Prof. Dr Walther EHALT. Ärztl Leiter des Arbeitsunfallkrankenhauses Graz der Allgemeinen Unfallversicherungsanstalt. Unter Mitarbeit von Georg Gelehrter, Josef Riess, Otto Schmid-Schmidsfelden, Alois Titze, Fritz Werkgartner und Herbert Kaloud. 9½×6½ in. Pp. xii+432, with 2 farbbildern and 637 figures in 249 illustrations, and 30 tables. 1961. Stuttgart: Ferdinand Enke Verlag. Price Geheftet DM 90; Ganzleinen DM 94.

Those of us who know Ehalt and recognise his thoroughness and commonsense will not be surprised to find the same qualities in this excellent book. Clear, thorough and informative it leaves little to a reviewer but to recommend it. The first half of the volume is concerned with general principles, and those who possess a standard text-book of fractures may well start reading the special part of the volume on page 206.

In the treatment of tetanus Austrian surgeons have the advantage of dealing with a population which is actively immunised. How much simpler the life of a casualty officer in England would be if the same advantages were extended to the whole population here! Not only has the casualty officer the invidious decision whether A.T.S. should be given or not, but he should also arrange for active immunisation of the patient so that a subsequent injury can be treated with toxoid. It is true that the

extent of deaths from tetanus in this country is extremely small, but it would be a small price to pay for the peace of mind of both doctors and patients if legislation to ensure active immunisation of the population were passed.

Fractures of the lateral epicondyle of the humerus are clearly divided into displaced and undisplaced. Emphasis is laid on the fact that displaced fractures need open operation. Unfortunately attention is not drawn to the fact that clinical evidence of union of this fracture is inadequate and that radiological evidence of union must be obtained as continued observation of these cases shows that a large number fail to unite and subsequently lead to deformity of the elbow joint.

Although a shortening of 1·5 centimetres can be tolerated in fractures of the femoral shaft, fixation by wire through the tibial tuberosity is recommended in children up to the age of two, and no mention can be found of the simple and reasonably efficient treatment of such fractures by Bryant's method.

The illustrations and systematisation of the book are excellent. The treatment of burns and fractures of the skull are included. The volume is dedicated to Lorenz Böhler in honour of his seventy-fifth birthday. It is a worthy monument.—J. G. BONNIN.

Knochenstrüktur als Verbundbau, Versuch Einer Technischen Deutung der Materialstruktur des Knochens. By Prof. Dr Karl-Heinrich KNESE. $10\frac{1}{2} \times 7\frac{1}{2}$ in. Pp. 56, with 26 figures and 4 tables. Index. 1958. Stuttgart: George Thieme Verlag. Kartoniert DM 19.50.

The architectural units from which bones are built have been subjects of interest and importance to bone histologists and orthopaedic surgeons for a long while. One of the most interesting of these units is the " Haversian system " or " osteone," and the present monograph gives an account of the anatomy of these structures as seen in cortical bone—particularly in relation to the mechanical properties of bone tissue.

Clopton Havers, as long ago as 1691, described the vascular canals of compact bone: subsequent writers, particularly Todd and Bowman (1845) and Tomes and DeMorgan (1853), used the term " Haversian system " to describe the tubular collections of concentrically arranged bone lamellae surrounding each vascular canal. Tomes and DeMorgan—respectively a dental surgeon and a general surgeon at the Middlesex Hospital in London—knew that Haversian systems were formed by the deposition of successive lamellae of bone on the walls of cylindrical cavities—resorption cavities—in the tissue: they were able to show that, even in adult life, removal and formation of tissue were continually in progress in normal cortical bone, and that the histological structure of bone tissue was determined by these two processes. The term " osteone", which is much used in the continental literature, was introduced by the German anatomist, Biedermann, in 1914, to describe the " bone tubule " or " Haversian system " (*cf.* " nephron," " neurone," as applied to structural units of the kidney and the nervous system), and, in recent years, a considerable amount of information has been collected on the morphology and function of these structures.

During early life the central space of each osteone contains one or more capillary blood vessels, from which the nutrition of the cortical bone tissue is derived: in old age the central canals and their blood vessels undergo progressive obliteration, and extensive areas of bone tissue become metabolically inactive. Most osteones have an approximately longitudinal orientation in cortical bone, and this appears to be determined—at least in part—by the mechanical stresses to which a particular area of bone is subjected: reconstruction of individual osteones from serial sections (Cohen and Harris 1958) has shown that they actually follow a spiral course, and that they form communications between the periosteal and endosteal surfaces of the cortex. Individual osteones vary greatly in diameter and configuration, in the arrangement of their constituent lamellae, and in their degree of mineralisation. The microradiographic studies of Amprino and Engström (1952) first directed attention to this variation of mineral content: subsequent studies have shown that the growing or recently formed osteones are the least mineralised, and that there is an increase in mineral content with increasing age of the structure. Of particular interest have been studies with bone-seeking radio-isotopes such as ^{45}Ca or ^{90}Sr (Ponlot 1960) which show a relatively rapid exchange to occur between the circulating blood calcium and that present in the recently formed and still incompletely mineralised osteones. This rapid exchange is responsible for the uneven distribution of bone-seeking radio-isotopes after their injection into the body, these materials being deposited in greatest concentration in the osteones in process of formation and mineralisation at the time of administration. Work at present going on in a number of laboratories, using tetracyclines as vital markers of growing bone surfaces (Frost, Villanueva and Roth 1960), is making possible the determination of the rate at which bone is being deposited on the growing surfaces of developing osteones, and thus the rate of renewal—or " turnover "—of cortical bone tissue. From all these studies we are able to see how the histological structure of cortical bone gives an indication of the cellular processes which have shaped the tissue

in any given area. One of the central questions of bone physiology relates to the local control of these processes. We assume that the factors concerned are numerous and complex: we are completely ignorant of the mechanisms involved, and it may be that more detailed knowledge of the histological structure of cortical bone tissue will provide important clues in this direction.

The present work from the Institute of Anatomy of the University of Kiel (actually Volume 4 of Bargmann and Doerr's *Zwanglose Abhandlungen aus dem Gebiet der normalen und pathologischen Anatomie*. Stuttgart: Georg Thieme) discusses certain features of bone structure in relation to the mechanical function of the tissue. The author, in his previous writings on the subject (Knese, Ritschl and Voges 1954), has given us an account of the complexity and diversity of osteone structure and arrangement in cortical bone. Classifying individual osteones, as they appear in transverse sections, on the basis of their size, shape and obliquity, and on the structure and arrangement of their constituent lamellae, he has devised a complicated system of nomenclature and recording to demonstrate the way in which the osteone structure varies according to the bone concerned and the precise anatomical situation within the bone. Fascinating as these detailed studies are to the expert, it is not yet possible to interpret the results in any meaningful fashion. Why, for example, are the osteones of the central part of the cortex of a bone usually of greater diameter than those situated peripherally? Does this difference in size of osteones give the different areas of bone tissue any different physical or biological properties? We just do not know the answers to these questions. Neither do we know why certain osteones are made up of a finely fitted array of collagen fibres, while others consist of alternating lamellae of oriented fibres. Although Knese's work does not provide answers to such questions as these, it allows us to formulate the questions: he has documented in great detail the morphological characteristics of osteones as seen in frozen sections of decalcified bone when viewed by polarised light, in a variety of situations in the normal skeleton—particularly in the limb bones of one forty-three-year-old subject studied in considerable detail.

It is quite clear that the mechanical properties of bone tissue are related in some way to its histological structure. In the present work Knese suggests that the association of two dissimilar components—the traction-resistant collagen fibres and the compression-resistant mineral material—is the important structural feature from a mechanical point of view. In this connection he makes the apt comparison between the structure and mechanical properties of bone and those of reinforced concrete. He further argues, although on what evidence it is not clear, that the comparison can usefully be extended to " prestressed " concrete, his view being that the collagen fibres of bone are also prestressed and that this aids their resistance to compression. He then goes on to suggest that the structural complexity of bone, to which reference has already been made, may cause this tissue to have different mechanical properties in different situations. If this is the case it should be possible to demonstrate such local variations, and it would be important to know what they are.

Not only does Knese adopt a complicated scheme of description and classification of osteones, but, in common with other German authors (Rollet 1871, Petersen 1930), he uses a scheme of " order stages " to describe certain general aspects of bone structure. To the present reviewer this appears to be complicated and unnecessary. It is clear that a particular bone, such as the femur, is an aggregate of smaller and possibly simpler structural units, but one wonders what is to be gained by describing the arrangement of compact and spongy bone as " structures of the first order," and passing downwards to collagen fibres and bone crystallites, which are regarded as the ultimate units of structure " of the fifth order." The intermediate " orders " are particularly artificial: structures " of the fourth order " are lamellae; structures " of the third order " are " lamellar systems," and thus include osteones; structures " of the second order " are " specific topographical features of lamellar systems." Surely the anatomical configuration of the tissue and its functional implications can be described without this hierarchial organisation.

Anyone who is personally concerned with histological studies of bone structure will be interested in Knese's work—interested despite its complexity, some, but not all, of which is a result of the complexity of the biological material itself. The technical preparations and the photomicrographs illustrating them are elegant and informative, and the production of the book is excellent. The results of the careful and very laborious work described are all raw material for some general theory of bone structure, but no coherent explanation yet emerges from them. Perhaps Professor Knese will one day be able to produce such a theory for us—and what an exciting day that will be.—H. A. SISSONS.

REFERENCES

AMPRINO, R., and ENGSTRÖM, A. (1952): Studies on X Ray Absorption and Diffraction of Bone Tissue. *Acta Anatomica*, **15**, 1.

BIEDERMANN, W. (1914): In *Handbuch der vergleichenden Physiologie*. Herausgegeben von H. Winterstein. Jena: G. Fischer.

COHEN, J., and HARRIS, W. H. (1958): The Three-Dimensional Anatomy of Haversian Systems. *Journal of Bone and Joint Surgery*, **40-A**, 419.

FROST, H. M., VILLANUEVA, A. R., and ROTH, H. (1960): Measurement of Bone Formation in a 57 Year Old Man by Means of Tetracyclines. *Henry Ford Hospital Medical Bulletin*, **8**, 239.

HAVERS, C. (1691): *Osteologia Nova, or Some New Observations of the Bones, and the Parts Belonging to Them, with the Manner of their Accretion and Nutrition*, p. 294. London: S. Smith.

KNESE, K. H., RITSCHL, I., and VOGES, D. (1954): Quantitative Untersuchung der Osteonverteilung im Extremitätenskelet eines 43 jährigen Mannes. *Zeitschrift für Zellforschung und Mikroskopische Anatomie*, **40**, 519.

KNESE, K. H., VOGES, D., and RITSCHL, I. (1954): Untersuchungen über die Osteon- und Lamellenformen im Extremitätenskelet des Erwachsenen. *Zeitschrift für Zellforschung und Mikroskopische Anatomie*, **40**, 323.

PETERSEN, H. (1930): Die Organe des Skelettsystems. In Moellendorff's *Handbuch der mikroskopische Anatomie des Menschen*. Band 2. Teil 1. Berlin: J. Springer.

PONLOT, R. (1960): *Le radiocalcium dans l'étude des os*. Brussels: Éditions Arscia. Paris: Masson et Cie.

ROLLETT, A. (1871): In *Handbuch der Lehre von den Geweben des Menschen und der Thiere*. Herausgegeben von S. Stricker. Leipzig: Wilhelm Engelmann.

TODD, R. B., and BOWMAN, W. (1845): *The Physiological Anatomy and Physiology of Man*. London: John W. Parker.

TOMES, J., and DEMORGAN, C. (1853): Observations on the Structure and Development of Bone. *Philosophical Transactions of the Royal Society of London*, **143**, 109.

Biophysical Investigations of Bone Transplants and Bone Implants. An Experimental Study. By Kaj HOLMSTRAND. 9×6 in. Pp. 66, with 34 figures and 2 tables. 1957. Copenhagen: Ejnar Munksgaard. Acta Orthopaedica Scandinavica, Supplement 26.

Biophysical Investigations of the Mineral Phase in Healing Fractures. By Ulf NILSONNE. $9\frac{1}{2} \times 6\frac{3}{4}$ in. Pp. 80, with 27 figures and 4 tables. 1959. Copenhagen: Ejnar Munksgaard. Acta Orthopaedica Scandinavica, Supplement 37. Price Da. kr. 20.

These two short monographs, from the Department of Medical Physics of the Karolinska Institute in Stockholm, describe the application of certain biophysical techniques (particularly microradiography, x-ray diffraction, and autoradiography) to bone tissue.

Microradiography involves the preparation of an x-ray picture of a thin section of undecalcified bone on a high-resolution photographic emulsion. A number of interesting and important studies by Professor Engstrom and his colleagues in Stockholm, as well as by workers in other centres, have established that such a picture shows the distribution of the mineral material (hydroxyapatite) of the bone and that, under certain specified conditions, it is possible to measure the amount of such mineral material present in any particular area of the microscopic preparation. Before this, the only information on the mineral content of bone tissue was obtained by chemical analysis, and this could be applied only to much larger areas of tissue. Microradiographic studies of normal bone have shown appreciable variation in the degree of calcification of different areas of tissue, and these can be explained by a progressive increase in the calcification of bone tissue—in particular, the developing osteones of cortical bone—as the tissue matures. In Holmstrand's work, determinations of mineral content in a series of autografts and homografts in rabbits showed a similar increase in the degree of mineralisation of the reactive bone tissue surrounding the transplant. This was observed to occur over a period of sixty weeks, and took place at a greater rate in the first few weeks than in the remainder of the period. The average increase in mineralisation was about 2 per cent in ten weeks, and the average time to achieve " full " mineralisation of the reactive bone was estimated at ninety weeks. In Nilsonne's work the same type of observation was made on the bony callus tissue in experimental fractures in rats and dogs, and a similar increase in the degree of mineralisation was observed. Again, mineralisation proceeded most rapidly in the first weeks after the formation of the callus tissue. The results indicate, then, that this slow increase in the degree of mineralisation of newly formed bone structures is probably a feature of all types of bone tissue. The technique is one which will doubtless be applied to the study of many other aspects of bone mineralisation.

Another procedure used in each of the studies is micro x-ray diffraction. A narrow beam of monochromatic x-rays is directed perpendicular to a particular part of the bone section. In passing through it the x-ray beam is diffracted and, if the crystal units of the bone mineral are randomly oriented, the reflections from the crystal planes will show up as a set of concentric rings whose spacing is characteristic of the material concerned. If the crystal units are oriented in a particular direction the x-ray diffraction pattern will show this by intensification of the rings over certain arcs. A somewhat different use of x-ray diffraction allows determination of the size of the crystal units. Both in the reactive bone surrounding grafts, and in fracture callus, the diffraction patterns obtained were

characteristic of hydroxyapatite. In newly formed tissue, there was no indication of any orientation of the crystal units, but later such orientation became evident. From a study of the reactive bone surrounding grafts, Holmstrand reached the conclusion that the orientation of the bone crystallites is independent of the mechanical forces acting on the tissue. From the study of fracture callus, Nilsonne concluded that the crystallites became oriented predominantly in the longitudinal axis of the bone concerned: this corresponded to the longitudinal orientation of the callus tissue on the histological level, but no observations were made on the relationship to mechanical forces. This interesting question concerning the effect of mechanical forces on the orientation of the bone crystallites, and the relationship between this orientation and that of the larger histological units of the tissue, would seem to justify further work.

Another biophysical technique employed in these studies is autoradiography after the injection of radioactive strontium (Sr^{90}). The distribution of radioactive material in a bone section is demonstrated by placing this in contact with a photographic emulsion: sites of activity show as localised areas of blackening when the emulsion is developed. In both graft material and fracture repair the newly formed bone tissue showed heavy uptake in comparison with mature bone: the greatest uptake is in the least mineralised tissue.—H. A. SISSONS.

Le Radiocalcium dans l'Étude des Os. By R. PONLOT, Chercheur agréé d'I.I.S.N. 9×6 in. Pp. xvii+213, with 86 figures, 2 tables and appendix. Index. 1959. Bruxelles: Éditions Arscia. Paris: Masson et Cie. Price 46 Fr. N.S.

Autoradiography following the injection of radioactive isotopes is now widely used in the study of bone. Precise interpretation of the autoradiographic image produced by the tissue at different stages of development and at different times after injection is an essential part of this technique. In this monograph Dr Ponlot—working in the laboratory of Professor Lacroix—combines a number of the contributions already made in this field with some important new observations on the localisation of 45Ca in bone.

He first studies, in considerable detail, the initial site of calcium deposition in the osteone of adult bone, namely the border-line (" ligne-frontiere ") between the pre-osseous layer (" lisere pre-osseux ") and the heavily calcified tissue which surrounds it. By means of a variety of histochemical and histological techniques not previously used at this site, he establishes the peculiar properties of this zone. He demonstrates a change in the ground substance coincident with calcification. He challenges the exponents of the theory that collagen alone is important to the initiation of calcification by showing (with 3H labelled glycine) that collagen is present throughout the pre-osseous layer and that it is formed two to four days before the deposition of calcium. The term " lisere pre-osseux " is suggested by the author to distinguish the border of uncalcified tissue present in normal Haversian systems from the abnormally wide " osteoid-border " seen in rickets.

A detailed analysis of autoradiographic imprints follows. The importance of a system of double reference to these is stressed. Thus the autoradiograph should always be compared with the section itself stained to show the pre-osseous layer, and with the microradiograph. The use of autoradiography in studying the pattern of growth of a long bone is demonstrated.

Finally the concept of " an osteone " is reappraised. Dr Ponlot has shown the formation of osteones along the periosteal surface of long bones wherever rapid growth in the diameter of the shaft occurs. These osteones resemble those formed in resorption cavities in all respects except in their site of origin and in the absence from the periosteal osteones of a true cement line. It is suggested that the term " osteones d'addition " be adopted for the periosteal osteones in order to distinguish them from " osteones de remplacement," i.e., those formed as a result of remodelling.

It is regrettable that the monograph, which is written in French, is not summarised in any other languages. All the techniques used are fully described and the text is excellently illustrated. Many more topics emerge than can be hinted at in a short review, and the whole is a refreshing and stimulating approach to fundamental problems of bone structure and function.—Lorna SECKER WALKER.

The Surgical Anatomy and Pathology of the Supraspinous and Interspinous Ligaments of the Lumbar Spine with Special Reference to Ligament Ruptures. By Pentti M. RISSANEN. 9×6 in. Pp. 100, with 57 figures. 1960. Acta Orthopaedica Scandinavica, Supplement 46. Copenhagen: Ejnar Munksgaard. Price Dan. kr. 18.

This monograph is based on the macroscopic and microscopic study of 306 normal cadaveric lumbar spines from those of all ages. After introductory sections explaining the aims of the investigation, and the material and methods employed, the first major section deals with the supraspinous ligament. The most important findings were, firstly, that contrary to accepted teaching, the ligament did not extend throughout the whole length of the lumbo-dorsal spine, but ended at either the third or fourth

lumbar vertebra, and secondly that with ageing, fibrocartilaginous metaplasia took place, starting in the deeper layers, often progressing later to ossification.

The remainder of the work deals with the interspinous ligaments. The first section, a study of the appearance in the foetus and infant, showed that the ligament develops concurrently with the spinous processes. This is followed by a detailed investigation into its structure, the main findings of which were in the upper three lumbar interspaces; the ligament could be subdivided into four fairly constant layers from before backwards, whereas, in the lower two spaces, only three layers could be distinguished, the fibres of which varied widely in direction and strength.

The next section deals with age changes, and, over twenty years old, degeneration was invariably found in the tendinous tissue of the ligaments. Later, cavities occurred, often microscopically resembling joints. The final section discusses ruptures in the interspinous ligament, which were formed in 21 per cent of subjects over twenty years of age. In the large majority the ruptures were located in the lowest two spaces, which are also the levels in which intervertebral disc prolapse is most common.

The monograph is purely an anatomical study; no attempt being made to correlate the various findings with any clinical picture. It is a painstaking study of structures whose importance as a cause of low back pain may not be sufficiently appreciated and will be of value to those interested in the subject.—J. N. ASTON.

Lumbar Intradiscal Pressure. Experimental Studies on Post-mortem Material. By Alf NACHEMSON. $9\frac{1}{2} \times 6\frac{1}{2}$ in. Pp. 104, with 58 figures and 35 tables. 1960. Acta Orthopaedica Scandinavica, Supplement 43. Copenhagen: Ejnar Munksgaard. Price Dan. kr. 18.

This is an interesting work on the measurement of " intradiscal pressures " in cadaveric disc specimens by means of an electronic device which might some day be developed for use *in vivo*. To avoid the disadvantages of injecting fluids into the disc with an open-ended needle, as used by previous workers, the author has used a needle with a lateral opening covered by a sleeve of polythene tubing to act as a diaphragm. A number of technical difficulties face the transmission of the deflections of this diaphragm through the water-filled needle to a pressure transducer, and these difficulties, of which the most erratic was the inconstant elastic behaviour of the polythene diaphragm, are described.

The principal findings are, firstly, that the nucleus pulposus behaves as a hydraulic medium when loads are imposed on the intact disc, and this applies even when there is considerable degeneration. When degeneration is so severe as to produce cavities, the nucleus no longer behaves hydraulically as might be expected. Hydraulic behaviour is indicated by the record of pressure inside the nucleus remaining constant no matter what way the recording diaphragm of the needle is directed.

Secondly, it is shown that the " intradiscal " pressure is 30 to 50 per cent higher than what would be calculated from a known load on the area of the disc. In other words, the normal disc takes more load on to the nucleus than it does on the annulus, which implies that the vertical stress in the annulus is less than the tangential stress. These findings do not apply to the severely degenerate disc where the annulus takes a much higher load.

One aspect of this monograph which this reviewer feels should be criticised is the inclusion, without adequate discussion, of the part played by the posterior intervertebral joints and pedicles in the transmission of load. It is recorded that removal of the posterior elements caused a rise in intradiscal pressure of about 20 per cent, which implies that the posterior elements were transmitting this amount of weight. No attempt is made to explore further the mechanism of this unlikely happening, which is all the more exasperating since Nachemson was part author of a previous work which suggested that the posterior elements of the spine were not involved in the transmission of load. It is possible that the posterior elements may transmit load in extension but it seems impossible in the neutral or flexed position of the spine. This is an example of an observation's being afforded the dignity of publication when in conflict with commonsense; unless a researcher takes an investigation far enough to explain an apparent inconsistency, or far enough to show that he has eliminated all known experimental errors, its appearance in the literature merely confuses the issue.

Despite this reservation, the monograph is valuable and interesting, and will be helpful to others in a field of research which is only just starting.—John CHARNLEY.

Die Versorgung von Ohnhändern. By Prof. Dr Otto Boos, Oberarzt der Orthopädischen Universitätsklinik und Poliklinik, Tübingen. Mit einem Beitrag von Dr med. Ernst GOETZ, Ob. Reg. Med. Rat in der Ärztlichen Abteilung des Bundesministeriums für Arbeit und Sozialordnung, Bonn; und einem Geleitwort von Prof. Dr Lothar KREUZ, Tübingen. $9\frac{1}{2} \times 7$ in. Pp. 151, with 33 figures (some in colour) and 14 tables. 1960. Stuttgart: Friedrich-Karl Schattauer-Verlag. Price DM 22.

This paper-backed book written in German deals with the treatment and after-care of people who lack all the digits of both upper limbs whatever the level of the defect.

The writer, Professor Boos, draws on his experience in World War II of 263 cases treated at the Oskar-Helene-Heim Clinic in Berlin, an institution which seems to combine some of the features of Roehampton. When he moved to Tübingen Professor Boos reviewed all the patients in south-west Germany—108 in number—who had been treated by Krukenberg's method of converting the forearm stump into a two-pronged claw. He reviews the prosthetic methods of cineplastic procedure of the upper arm stump and of the phalangisation of the metacarpals, and then deals with his favourite subject and gives a full description of the Krukenberg procedure, the difficulties which are encountered and the complications. He also deals with the subsequent treatment of cases based on the phantom hand in which muscle re-education is employed. He suggests that it is important for each patient to be trained for his daily life, the ultimate training to take into consideration the future occupation of the patient. About 58 per cent of patients without hands are employed. The amputees are generally fitted with a prosthesis which is activated by the prongs, and this was Krukenberg's original purpose, but most of these patients find that the bare stumps are more useful. They, however, need specially modelled handles for most objects they use.

A section is devoted to the great problem of training blind people who have lost their hands.

The book goes on to deal with the social as well as the surgical problems encountered. The author says he advised patients in all matters, even in the choice of their wives. Most of these people do marry. This is a point of great significance when it is considered that nearly half of these patients need to be helped with their toilet, and only slightly fewer with their washing.

The author considers that the treatment of the handless should be concentrated in special centres where the necessary arrangements can be made so that surgeons can gain enough cumulative experience.

Fortunately the loss of both hands rarely occurs in civilian life and one cannot but hope that the unique experience gained by this author and others will not need to be repeated.—Leon GILLIS.

Transactions of the International Society of Plastic Surgeons. Second Congress, London, 1959. Edited by A. B. WALLACE, M.Sc., F.R.C.S.Ed., Editor of the *British Journal of Plastic Surgery*. 10×7½ in. Pp. xxiv+589, with many illustrations. Index. 1960. Edinburgh and London: E. & S. Livingstone Ltd. Price 90s.

Plastic and orthopaedic surgery are fields which have much in common and it is to be expected that in this volume, *Transactions of the Second Congress of the International Society of Plastic Surgeons*, there will be much to interest the orthopaedic surgeon.

Some eighty papers, embracing all aspects of reconstructive surgery, are reproduced in full, together with brief summaries in French, Spanish and German.

The papers are grouped in sixteen sections, chiefly according to the anatomical region discussed: for example—jaws, grafts for defects of facial bones and skull, genitalia, etc. Clearly this is no text-book and could be better described as a massive edition of a *Journal of Plastic Surgery*. As such it provides an interesting synopsis of advances, endeavours and problems in this expanding branch of surgery.

Some sections will be of particular interest to orthopaedic surgeons. There are eleven papers on various aspects of hand injury, and among these Littler's description of his neurovascular skin island transfer is outstanding. Harrison describes the retention of one slip of the intact sublimis with grafting for a divided profundus tendon. A novel means of splinting a finger in flexion is described by Planas of Barcelona, who inserts a simple stitch through the free border of the thumb nail and the nail of the finger to be held flexed.

Among several papers on tissue preservation, a preliminary report by Gibson of a living homograft cartilage bank with a rapid test for viability is of particular interest.

One might criticise the volume in that it lacks balance, that too much space is devoted to cleft lip and palate and not enough to the numerically greater problems of accidental trauma and burns. This is, however, the report of a Congress and its make-up was thus predetermined by the time allocated to the various subjects discussed.

The book is expensive at £4, 10s. but it is lavishly produced and the illustrations are excellent. It will interest all who undertake the surgery of repair.—R. L. B. BEARE.

Symposium sur l'Os Hétéroplastique. Under the Direction of Professor M. GUILLEMINET, Clinique de Chirurgie Orthopédique de la Faculté de Médecine de Lyon Hôpital Edouard-Herriot. 9½×7 in. Pp. 103, with 64 figures and 12 tables. 1959. Revue de Chirurgie Orthopédique et Réparatrice de l'Appareil Moteur, Volume 45, Supplement No. 1. Paris: Masson & Cie. Price $1,65.

In this country bone heterografts have never been widely used. This symposium which records experiences with this material in several continental centres is therefore of great importance to us.

In Lyon, where this meeting was held, facilities have been developed for obtaining sterile calf bone, which is preserved either by refrigeration at −30 degrees Centigrade, or more recently by freeze drying. No less than 1,500 grafts have been distributed to surgeons in the past ten years.

Several of the papers describe attempts to assess this material experimentally. While they clearly show that heterografts can behave like autograft or homograft, the process is usually delayed; sometimes greatly.

The papers on the clinical use of heterografts are very difficult to evaluate because of the variety of techniques and operations described. In general it is evident that, although brilliant results can be obtained, a high proportion of failures must be expected. Complications such as total rejection of the graft and persistent discharging sinuses are frequently mentioned. Marion and his colleagues had a success in only 51 per cent of cases. Desbrosses reports success in 90 per cent when using heterograft to fill bone cavities, but his illustrations do not support his text.

On the basis of the evidence presented in this symposium bone heterograft would appear to be inferior to both autograft and homograft.

The enthusiasm and industry with which Professor Guilleminet and his colleagues have pioneered this field command respect, and the results of their further work with freeze dried heterograft will be watched with interest.—J. CHALMERS.

Progressi in Ortopedia. Scritti Medici in Onore di UGO CAMERA. Nel 50° Anno Della Sua Attività Scientifica e Professionale 1909–1959. A cura di C. A. LIÈVRE. Aiuto nell'Istituto Chirurgico Ortopedico "Regina Maria Adelaide" Clinica Ortopedica dell'Università—Torino. 11½×8¼ in. Pp. 553, with many figures. 1959. Torino: Edizioni Minerva Medica. Price L. 8,500.

This is a compilation of unrelated works by a large number of authors from many countries to honour a distinguished colleague. There are about one hundred such contributors to the present volume, knowing that for the most part their contributions would be as good as lost to the ordinary medical reader. This is because, up to the present time, no way has been found to list the works appearing in these volumes either by authors or by subject matter in the indexes of bibliography. Perhaps it is for this that the individual contributions as a homage are sent in, for they will be sacrificed at the altar of the god of friendship.

The quality of many of the works in the present compilation, and their number, are evidence of the high consideration and affection that so many orthopaedic surgeons have for Hugo Camera. His personal contribution as a pioneer in many fields of the speciality gave Camera a place in Italian orthopaedics side by side with Putti, Delitala and others from the past generation. He was the first to advocate the shortening of the longer limb to treat disparity in growth and it was also he who pointed out first that coxa vara could be the result of an injury of the cartilage anlage.

It would be impossible to name some of the papers published in this volume without doing injustice to others and for this reason I abstain, but for those who can read Italian it is worth perusing its index where they will find a large area of the field of orthopaedics covered by men whose names are at the forefront of continental orthopaedic surgery. It consists of 553 well printed pages and is profusely illustrated.—Joseph TRUETA.

Chemical Osteosynthesis in Orthopaedic Surgery. By Michael P. MANDARINO, M.D., Associate, Department of Orthopaedic Surgery, Hahnemann Medical College and Hospital; Associate, Department of Orthopaedic Surgery, Misericordia Hospital, Philadelphia, Pennsylvania. American Lecture Series No. 391. 9¼×6½ in. Pp. x+72, with 36 figures. Index. 1960. Springfield, Illinois: Charles C. Thomas, Publisher. Oxford: Blackwell Scientific Publications Ltd. Toronto: The Ryerson Press. Price 36s.

In the first fourteen pages of the text many examples are given of isocyanate and di-isocyanate reactions which can take place in the formation of a polyurethane resin. The author does not, however, give the constituents used in the manufacture of the polyurethane, marketed under the trade name Ostamer and about which the book is written. In a polymerisation reaction there is never complete reaction of the constituents. In the case of Ostamer, the theoretical degree of polymerisation is the more difficult to achieve at operation because of the method of mixing the ingredients, the lack of control of the temperature of the reaction and the possible inhibitory effects of blood and fat. A list of mechanical properties is given which presumably has been obtained using specimens of materials prepared under laboratory conditions. Ostamer can be likened to a cement which can be used to fill bone defects and repair fractures. Like cement, the resin has a high compression strength—20,000

pounds/square inch, but a low tensile strength—800 pounds/square inch. When Ostamer is used in the treatment of long bone fractures of the lower limb, an intramedullary rod reinforcement should be incorporated. Bone chips and anorganic bone may be added to the resin, but these materials reduce its mechanical properties. From a study of the tissue reaction in various animals it would appear that, with the material tested, no undesirable tissue reaction resulted.

The author gives some details of fifty cases which he has treated. In some of these cases there had been previous bone infection, yet union of the fractures was achieved. Of the fifty cases, twenty-eight healed uneventfully, five continued to drain and three tibial cases were failures. It is difficult to ascertain from the text the fate of the other fourteen.

No one would deny that there is a place for a gap-filling cement in orthopaedic surgery which is apparently resorbed and replaced by bone, but its use must be limited until more is known about the long-term tissue reaction and route of excretion of polyurethane resins in man. As the author says: " The first indication for Ostamer fixation at surgery, therefore, would seem to be the non-union of bone in which previous surgery had failed."

This book lacks the details which are necessary for the accurate assessment of a technique.—John T. SCALES.

Wirbelgelenk und Bandscheibe. Ihre Beziehung zum vertebragenen Schmerz Zugleich eine Stellungnahme zur Chiropraktik und Begutachtung. By Prof. Dr L. ZUKSCHWERDT, Dr med. habil. E. EMMINGER, Dr F. BIEDERMANN, and Dr H. ZETTL. Zweite, neu bearbeitete und erweiterte Auflage. $10\frac{1}{2} \times 7\frac{1}{2}$ in. Pp. 319, with 160 figures. Index. 1960. Stuttgart: Hippokrates-Verlag. Price DM 58.

This is an important book. It makes a sincere effort to bridge the gap between the practising orthopaedic surgeon, the pathologist and the chiropractor or manipulative surgeon.

It starts from two bases: the recognised beneficial effect of manipulation, and the ignorance of the special pathology that might be benefited by manipulation. The authors contend that the work of Schmorl and Junghanns has largely been directed to studying the pathology of the intervertebral disc and has left the pathology of the lesser intervertebral joint almost untouched.

The first section of the book is devoted to the re-examination of disc pathology and a study of the pathological changes in the lesser intervertebral joints, together with the possible causes of narrowing of the intervertebral foramina. These include not only disc prolapse, but disc degeneration with bony peripheral hypertrophy, hypertrophy of the ligamentum flavum, formation of the fatty bodies around the lesser intervertebral joints, arthritic changes in the lesser intervertebral joints and anoxaemia of the issuing nerve root as a result of the vascular pathology. The almost universal presence of a small intra-articular disc in the lesser intervertebral joint is clearly demonstrated, and it is suggested that acute lumbago is due to nipping of this like a medial semilunar cartilage [medial meniscus]. The variety of possible pathology in narrowing intervertebral foramina is equalled by the variety of treatments to which a cure has been attributed but which do not always act with certainty. A careful examination of the pathology of spondylosis deformans and its relation to back pain is followed by a routine section on the signs and symptoms of nerve root irritation on an anatomical basis. This is perhaps the only part of the book with which there will be general agreement.

The most valuable section now follows: a description of the varieties of pain related, or possibly related, to spinal disease. Here the authors go further than usual by attributing a wider range of peripheral disorders to nerve root irritation than is generally accepted, though this hypothesis forms the basis of chiropraxis. Thus, cervical migraine, peritendinitis of the shoulder, tennis elbow, nocturnal paresthesia and dysphagia are all considered as well as the more familiar syndromes with an agreed vertebral origin.

The borders of the known and the unknown are carefully examined and the deficiencies in knowledge admitted, but this book succeeds in offering a more rational basis for manipulative treatment than previous volumes. While the value of surgical treatment is not denied, the difficulties in selecting suitable cases are clearly stated, and the well-known fact that the expected pathology is not always found at operation and yet the symptoms clear up is well considered. More attention, however, is devoted to routine conservative treatment in relation to the suggested pathology that varying degrees of nerve root compression are the basic cause of most of the syndromes described: pain, muscle spasm, sensory and muscular dysfunction and paresis, and sympathetic nerve disturbance. The manipulative methods employed are clearly described and illustrated.

This is an excellent review of the borders of the known and unknown in spinal pathology and, if a little biased towards manipulative treatment, is perhaps all the more stimulating.—J. G. BONNIN.

A Study of Home Accidents in Aberdeen: an investigation financed by the Nuffield Provincial Hospitals Trust. By Ian A. G. MACQUEEN, M.D., D.P.H., Medical Officer of Health, City of Aberdeen, and Lecturer in Public Health, University of Aberdeen. $8\frac{3}{4} \times 5\frac{1}{2}$ in. Pp. viii+100, with 10 figures, 2 appendices and 20 tables. Index. 1960. Edinburgh and London: E. & S. Livingstone Ltd. Price 12s. 6d.

Accidents in the home exceed even those on the road, and therefore deserve serious study. This book presents the work of a team. All domestic accidents in Aberdeen receiving medical attention during two years were recorded in relation to their nature, sex and age of patient, time and day of week, housing conditions and many other circumstances. In general, the conclusions are much as might be expected, with emphasis on the great susceptibility of children between one and four years old and of the elderly. There are several puzzling surprises, notably that old people, though specially liable to falls, were not unduly liable to such things as scalds, which were important in the young. Highly polished floors were probably the commonest single cause of domestic accidents. The conclusions may be a little unreliable for other cities, because Aberdeen has a serious housing problem, even by Scottish standards, and because it seems likely that the incidence and kinds of accidents had been influenced by a preceding educational campaign for their prevention. It is perhaps characteristic of Aberdonian reticence that alcohol is nowhere mentioned. Even in countries whose wine is not whisky, alcohol makes a considerable contribution to domestic accidents in every walk of life, often quite unobtrusively; but possibly assessment would be too difficult.—H. Jackson BURROWS.

Zur Frage der Unfall- und Berufsbedingten Sehnenscheidentuberkulose. By Prof. Dr Med. T. BURCKHART, Chirurgische Universitätsklinik Mainz. Direktor: Prof. Dr G. Brandt. 9×6 in. Pp. 24, with 2 figures and 6 tables. 1959. Hefte Zur Unfallheilkunde, Hefte 61. Berlin, Göttingen and Heidelberg: Springer-Verlag. Price DM 5.

As the incidence of tendon sheath tuberculosis from infection with human type of tubercle bacillus diminishes, accidental infection of the tendon sheaths as a result of occupational injury becomes relatively more common. In approximately a quarter of a million cases of injury to Union members employed in butchery, sixty-two cases of tendon sheath tuberculosis were found. In six cases the diagnosis was mistaken; in three the infection resulted from spread of bone and joint tubercle; in twenty cases investigation led to a denial of their occupational origin; seven cases remained under review. Twenty-six cases were, therefore, left of proven occupational origin.

As tuberculous disease of the skin does not lead to tendon sheath infection, the only possibility of accidental infection of a tendon sheath is by direct inoculation with bovine tubercle as a result of a penetrating wound. As the wound is often trivial it is frequently not notified and considerable delay in the diagnosis of tendon sheath infection usually occurs. This adds greatly to the difficulty in relating the infection to the injury.

This pamphlet is primarily concerned with the diagnosis of occupational disease and does not discuss treatment. It is an important and valuable paper for those concerned with industrial health and must be borne in mind by doctors treating injuries arising in slaughterhouses. The main points for consideration after such injuries may be summarised as follows: 1) The diagnosis must be established by biopsy. 2) The occurrence of a related accident must be completely proved. 3) The injury must have been of such a kind as to penetrate to the tendon sheath. 4) The site of the wound and the sheath infected must be in relation to each other. 5) The conditions of the accident must be such that contact with infected tuberculous material was probable and this is almost entirely limited to slaughterhouse work. Meat handling does not lead to infection. 6) The objective signs of tendon sheath tuberculosis must be established within a minimum of four weeks or a maximum of six months after the injury. 7) Typing of the bacillus must be carried out. The discovery of human type excludes an occupational origin. 8) Tuberculosis in other situations in the same patient makes the relationship to injury improbable.—J. G. BONNIN.

The Effect of Subperiosteally Implanted Autogenous Whole-thickness Skin Graft on Growing Bone. An Experimental Study. By Jaakko O. ELO. $9\frac{1}{4} \times 6\frac{1}{4}$ in. Pp. 90, with 56 figures and 17 tables. 1960. Acta Orthopaedica Scandinavica, Supplement 45. Copenhagen: Ejnar Munksgaard. Price Dan. kr. 18.

This monograph describes some experiments investigating the influence of subperiosteal implants of skin on the longitudinal growth of the rabbit's tibia.

Provided that an extensive stripping of periosteum was carried out at the time of implantation, a slight increase in length of the bone occurred. If the stripping extended to include the perichondrium of the epiphysial cartilage plate, the bone growth was diminished.

Skin can now be added to the long list of implants and irritants which when placed in the metaphysial region of a growing bone result in a small stimulation of growth. It does not appear to have any advantage that would recommend it for clinical trial.

A short but interesting review of the literature on the growth of long bones is a valuable part of this work.—J. CHALMERS.

Demonstrations of Physical Signs in Clinical Surgery. By Hamilton BAILEY, F.R.C.S.(Eng.), F.A.C.S., F.R.S.E., Emeritus Surgeon, Royal Northern Hospital, London; Consulting Surgeon, Italian Hospital; General Surgeon, Metropolitan Ear, Nose and Throat Hospital; Vice-President, International College of Surgeons; formerly Hunterian Professor, Royal College of Surgeons; and External Examiner in Surgery, University of Bristol. Thirteenth edition. 9×7 in. Pp. xvi+928, with 1,142 figures (many in colour). Index. 1960. Bristol: John Wright & Sons Ltd. Price 75s.

The thirteenth edition of this book is the last work of one who was perhaps the greatest surgical teacher of this generation, who died on March 26, 1961. The fact that there have been so many editions of this book—most of which have been reprinted—in the thirty-three years since it was first published is sufficient emphasis of its great popularity. The high standard of illustration and clarity of text are upheld in the new edition, and the price (75s.) is extremely modest.

It is always almost impossible to avoid enlarging any volume in repeated editions, and in this book the pages increased from 217 when it originally appeared to 456 in the last.

The present edition has had its scope deliberately widened, so that the book now has 928 pages, of which sixty compose the index. The text is thus four times that of the original, and over twice that of its immediate predecessor. Subjects of orthopaedic interest occupy about one-third of the volume.

The great increase in size means that the character of this work has changed—instead of being a small flat book which could be slipped into a student's brief case, and used at the bedside or ward rounds, it is now a tome comparing in size with many complete surgical text-books.

The new edition undoubtedly will have advantages for the postgraduate student, but how far these have been achieved at the expense of the undergraduate " to whom the book is still primarily addressed " time alone will show.—J. N. ASTON.

CONTRIBUTORS—MAY 1961

J. N. ASTON, Esq., F.R.C.S., 59 Harley Street, London, W.1, England.

Dr A. M. BARRETT, M.D., Department of Pathology, University of Cambridge, Cambridge, England.

R. L. B. BEARE, Esq., F.R.C.S., 149 Harley Street, London, W.1, England.

R. J. BERRY, Esq., M.A., Ph.D., Medical Research Council Experimental Genetics Research Unit, University College, Gower Street, London, W.C.1, England.

J. G. BONNIN, Esq., F.R.C.S., 51 Harley Street, London, W.1, England.

H. JACKSON BURROWS, Esq., M.D., F.R.C.S., 25 Upper Wimpole Street, London, W.1, England.

Dr R. A. CALDWELL, M.B., Ch.B., M.R.C.P.Ed., F.R.F.P.S.G., Department of Pathology, University of Sheffield, Sheffield, England.

Dr C. O. CARTER, M.R.C.P., Medical Research Council Clinical Genetics Research Unit, Institute of Child Health, Great Ormond Street, London, W.C.1, England.

JOHN CHALMERS, Esq., F.R.C.S., Orthopaedic Department, University of Edinburgh, 12 George Square, Edinburgh, Scotland.

JOHN CHARNLEY, Esq., F.R.C.S., 16 St John Street, Manchester 3, England.

The Rt. Hon. LORD COHEN OF BIRKENHEAD, M.D., F.R.C.P., Department of Medicine, The University, Liverpool 3, England.

Professor D. H. COLLINS, *O.B.E.*, M.D., F.R.C.P., Department of Pathology, University of Sheffield, Sheffield, England.

Dr M. DAMANSKI, M.D., Liverpool Regional Paraplegic Centre, Promenade Hospital, Southport, England.

T. J. FAIRBANK, Esq., F.R.C.S., Addenbrooke's Hospital, Cambridge, England.

Dr H. S. GILLESPIE, M.D., F.R.C.S.(C.), 402 Medical-Dental Building, 925 West Georgia Street, Vancouver 1, British Columbia, Canada.

LEON GILLIS, Esq., *M.B.E.*, M.Ch., F.R.C.S., Queen Mary's (Roehampton) Hospital, Roehampton, London, S.W.15, England.

M. A. GOODWIN, Esq., F.R.C.S.Ed., 494 Coniscliffe Road, Darlington, Co. Durham, England.

Dr R. HALL, M.R.C.P., Massachusetts General Hospital, Boston, Massachusetts, United States of America.

Prof. Dr OTAKAR HNĚVKOVSKÝ, Karlovo nám. 1, Prague 2, Czechoslovakia.

Dr R. W. HORNABROOK, M.D., M.R.A.C.P., 2nd Neurological Division, Bellvue Hospital, New York City, N.Y., United States of America.

R. NEVILLE HOULDING, Esq., F.R.C.S.Ed., African Life Building, Main Street, Bulawayo, Southern Rhodesia.

H. VAN DER HOUWEN, Esq., Anna Kliniek voor Orthopaedie, Leiden, Holland.

Dr BEN L. HULL, M.D., 1303 9th Street, Altoona, Pennsylvania, United States of America.

Dr L. M. JONCK, B.Ch.D., M.Sc., D.Sc., Department of Anatomy, Pretoria University, Pretoria, South Aftica.

Dr E. A. LENNON, M.C.R.A., F.F.R., 131 Wickham Terrace, Brisbane, Queensland, Australia.

P. S. LONDON, Esq., *M.B.E.*, F.R.C.S., Birmingham Accident Hospital, Bath Row, Birmingham 15, England.

A. T. MATHESON, Esq., F.R.C.S., P.O. Box 3289, Bulawayo, Southern Rhodesia.

S./Ldr. J. K. OYSTON, F.R.C.S., Royal Air Force Hospital, Cosford, nr. Wolverhampton, England.

ANDREW G. POLLEN, Esq., F.R.C.S., 129 The Vale, London, N.W.11, England.

Dr C. H. G. PRICE, M.D., Bristol Bone Tumour Register, Bristol Royal Hospital, General Hospital Branch, Guinea Street, Bristol 1, England.

NORMAN ROBERTS, Esq., F.R.C.S., 88 Rodney Street, Liverpool 1, England.

Dr J. T. SCALES, M.R.C.S., L.R.C.P., Department of Biomechanics and Surgical Materials, Royal National Orthopaedic Hospital, Brockley Hill, Stanmore, Middlesex, England.

Dr M. M. SCHECHTER, D.M.R.D., Neuroradiology Department, St Vincent's Hospital, New York City, N.Y., United States of America.

IAN K. SHARP, Esq., M.Ch.Orth., F.R.C.S., 37 Pulcroft Road, Hessle, Yorkshire, England.

Dr H. A. SISSONS, M.B., B.S., Institute of Orthopaedics, Royal National Orthopaedic Hospital, 234 Great Portland Street, London, W.1, England.

Dr JÖRGEN STOUGAARD, M.D., Amtssygehuset, Sorø, Denmark.

Dr A. TRIAS, M.D., Department of Orthopaedic Surgery, University of Chicago, 840 South Wood Street, Chicago, Illinois, United States of America.

Professor J. TRUETA, M.D., F.R.C.S., Nuffield Orthopaedic Centre, Oxford, England.

Mrs L. SECKER WALKER, Ph.D., Institute of Orthopaedics, Royal National Orthopaedic Hospital, 234 Great Portland Street, London, W.1, England.

EDRIC F. WILSON, Esq., F.R.C.S., 8 The Crescent, Plymouth, Devon, England.

Wing Commander C. B. WYNN PARRY, *M.B.E.*, D.M., M.R.C.P., D.Phys.Med., Medical Rehabilitation Unit, Royal Air Force, Chessington, Surrey, England.

The Journal of
Bone and Joint Surgery

EDITORIALS AND ANNOTATIONS

TENDON GRAFTS

In 1860 William Adams of London wrote: " Tendon is one of the few structures of the body, such as bone, cellular tissue, nerve tissue and blood vessels capable of reproduction or regeneration. The perfection of the process is in direct proportion to the absence of extravasated blood and inflammatory exudate. The sheath of the tendon, when consisting of loose textured areolar tissue, as in the tendo calcaneus, and other tendons surrounded by soft tissues, is of importance in preserving a connection between the divided extremities of the tendon; in furnishing the matrix in which the nucleated blastematous, or proper reparative, material is effused; and in giving definition and form to the newly developed tendinous tissue. The ultimate perfection of the reparative process is marred only by the adhesions of the deep surface of the new tendon to a greater or less extent with the neighbouring fibro-cellular tissue. These adhesions may limit the free play of the tendon, but will not interfere with sufficient motion being obtained."

Since these words were written, many investigations have been made into the nature of tendon healing and the behaviour of autogenous tendon grafts. The true facts concerning these processes in the human are not yet fully known, and even in carefully controlled animal experiments the results have been conflicting. Nevertheless, there is considerable confirmation of Adams's views.

Mason and Shearon (1932), while studying tendon healing in dogs, came to the conclusion that a graft maintains its vitality and unites to the host tendon by proliferation of the sheath tissue or the paratenon. They believed that this early union was completed by actual growth of the tendon cells (tenoblasts) from the ends of the tendon and the graft. Skoog and Persson (1954), working on rabbits, found that undifferentiated connective tissue arising from paratenon filled the gap between divided tendon ends and gradually matured to final union. They were unable to demonstrate any regenerative process arising from the tendon itself. They found that the cells in a free graft degenerate and that part, at least, of the graft is replaced by cells which infiltrate from the paratenon. Flynn, Wilson, Child and Graham (1960) confirmed the observation that union between host tendon and graft in dogs is effected mainly by proliferation of sheath or paratenon tissue, and found no evidence of tendon cell activity in the graft. On the contrary, they demonstrated complete ischaemic necrosis of the graft, which was subsequently invaded by tenoblasts from the host tendon until there was finally complete replacement of the grafted tendon. Lindsay and McDougall (1961) used the chicken's foot on account of the anatomical similarity between the digital flexor arrangement of chicken and man. They found that a graft survives as a whole, but although they demonstrated a process of repair consisting of revascularisation, increase in mature fibroblasts and gradual replacement or reconstitution of the original collagen, they found it impossible to determine the source of the cells which repopulated the graft.

Clinical experience supports the view of graft survival in man, because it is difficult otherwise to account for the rarity of rupture of the graft upon which considerable strain is placed long before reconstitution can occur. Peer (1955) took the view that the tendon cells

and stromal cells of a human graft survive, nourished by the rapid revascularisation which is known to occur. His opinion was based upon the examination of autogenous grafts buried in the abdominal fat. An active circulation was established within the graft in four to five days. Grafts were examined at intervals up to twenty-five days and after a lapse of seven and eight months. In no case could any evidence be found of invasion by fibroblasts from the surrounding tissue, and Peer concluded that the graft tendon cells must have survived as such unless they had been replaced by the fibroblasts in the connective tissue stroma of the graft.

An interesting and important observation emerging from the work of Skoog and McDougall is that any damage—even the passing of a suture and immediate withdrawal—to the epitenon (the thin adherent fibroblastic layer upon the tendon) leads to dense adhesions between tendon and surrounding tissues, thus restricting movement. Skoog, in particular, refutes the traditional view that it is the unsatisfied tendon ends which reach out to attach themselves to other structures. He calls attention to the common clinical observation that when a cut tendon end withdraws into an undamaged synovial sheath no significant adhesions are formed.

Brand's extensive experience in the use of free tendon grafts in the treatment of the paralytic hand of leprosy has led him to formulate certain rules for the operation of tendon grafting. The success of his operation for ulnar palsy has proved their value, but not all of these rules can be applied to the standard operation of graft replacement for divided flexor tendons in a digit. If they could be applied, the results would undoubtedly be improved. Even so, Brand's rules should be followed, whenever it is possible to do so, because they are in keeping with the experience gained from experimental and clinical work. It is fortunate that men like Brand and Riordan have shown how the principles and techniques of modern hand surgery can be applied to the treatment of this ancient but still widely prevalent disease. There are over ten million sufferers from leprosy in the world today and there is a great need for surgeons to become interested in this problem. They would find it a fascinating and rewarding task. Not only is there a vast amount of reparative surgery and research waiting to be done but there is also the need to train the leprosy patient in the care of the anaesthetic hand and foot and to help him back to his place in the community.

The hopes of many men, women and children rest on the orthopaedic and plastic surgeons of those countries where the disease is endemic. In the countries where there are no centres for reconstructuve surgery men trained in these concepts are needed to develop this work.

R. G. PULVERTAFT.

REFERENCES

ADAMS, W. (1860): *On the Reparative Process in Human Tendons.* London: John Churchill.

FLYNN, J. E., WILSON, J. T., CHILD, C. G., and GRAHAM, J. H. (1960): Heterogenous and Autogenous-Tendon Transplants. *Journal of Bone and Joint Surgery*, **42-A**, 91.

LINDSAY, W. K., and MCDOUGALL, E. P. (1961): Digital Flexor Tendons: An Experimental Study, Part III. *British Journal of Plastic Surgery*, **13**, 293.

MASON, M. L., and SHEARON, C. G. (1932): The Process of Tendon Repair. An Experimental Study of Tendon Suture and Tendon Graft. *Archives of Surgery*, **25**, 615.

PEER, L. A. (1955): *Transplantation of Tissues*, Vol. 1, p. 296. Baltimore: The Williams and Wilkins Company.

REPORT ON THE SCIENTIFIC MEETING ON REHABILITATION IN LEPROSY, VELLORE (1960): World Health Organisation. *Leprosy (Rehab.)*, 7.

SKOOG, T., and PERSSON, B. T. (1954): An Experimental Study of the Early Healing of Tendons. *Plastic and Reconstructive Surgery*, **13**, 384.